

MENTAL RETARDATION
ABSTRACTS

VOL. 7, NO. 4

OCTOBER-DECEMBER 1970

U. S. Department of Health, Education, and Welfare
Social and Rehabilitation Service
Rehabilitation Services Administration
Division of Mental Retardation
Washington, D. C. 20201

Mental Retardation Abstracts is a quarterly publication of the Division of Mental Retardation, Rehabilitation Services Administration. It is a specialized information service designed to assist the Division in meeting its obligation to plan, direct and coordinate a comprehensive nationwide program for those with mental retardation and related handicaps. Specifically, this service is intended to meet the needs of investigators and other workers in the field of mental retardation for rapid and comprehensive information about new developments and research results and to foster maximum utilization of these results.

Mental Retardation Abstracts is prepared under contract with the American Association for Mental Deficiency.

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Use of funds for printing this publication approved by the
Director of the Bureau of the Budget, June 30, 1969

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ACKNOWLEDGEMENTS

The publishers of the following journals have graciously granted permission to reprint abstracts and summaries:

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EDUCATIONAL AND PSYCHOLOGICAL SERVICES FOR THE MENTALLY RETARDED DEAF:
A SELECTED ANNOTATED BIBLIOGRAPHY

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This selected annotated bibliography relating specifically to the mentally retarded deaf focuses upon six areas of concern over a ten-year period with assessment, clinical characteristics, and educational training accounting for the majority of references. Within the past decade, there has been increased awareness of the problems involved in effective assessment, education, and planning for the multihandicapped mentally retarded population. Major problems in education for the retarded deaf center on qualifications and training of teachers, development of suitable curriculum and teaching techniques, and classroom grouping. Reliability and validity of diagnostic instruments are a major concern of investigators of assessment techniques for the mentally retarded deaf. As assessment procedures are refined, the number of individuals identified as mentally retarded deaf is expected to increase, thereby providing a substantial need for additional services for this population.

Overview

ANDERSON, R.M. Hearing impairment and mental retardation: A selected bibliography. *Volta Review*, 67(6):425-432, 1965.

A 100-item bibliography on hearing impairments in the MR is presented with reference to clinical and educational services.

ASSELSTINE, J. A future for the deaf retarded child. *Special Education in Canada*, 38(2):27-30, 1964.

Problems and statistical data related to deaf MR children are largely ignored. Coordination of services for the handicapped is needed. Diagnosis of hearing handicap and MR in a child is difficult and more valid testing procedures are needed.

COUNCIL FOR EXCEPTIONAL CHILDREN. *New Frontiers in Special Education*. Selected papers from the 43rd Annual CEC Convention, Portland, Oregon, April 20-24, 1965. Washington, D.C., Council for Exceptional Children, (No date), 298 p.

Ten papers relating to auditory impairment are presented.

DALE, D.M.C. *Deaf Children at Home and at School*. Springfield, Illinois, Charles C. Thomas, 1967, 272 p.

Deaf children are aided by good social training, lip-reading, and hearing aids, and IQ scores are not considered to be valid. Audiology clinics, preschools, regional education, and tutoring are needed, and programs should involve interaction with hearing children.

GOLDIE, L. The psychiatry of the handicapped family. *Developmental Medicine and Child Neurology*, 8(4):456-462, 1966.

Treatment of the family of a handicapped child should consider the problems of a "handicapped family."

LEENHOUTS, M.U. Problems accompanying children who are deaf and mentally retarded. In: Feisfeld, I.S., ed. *A Handbook of Readings in Education of the Deaf and Post School Implications*. Springfield, Illinois, Charles C. Thomas, 1967, 210-216.

Problems of MR deaf children enrolled in a residential school for the deaf involved class placement, curriculum, adept teachers, social adjustment, and residential living. Placement of MRs in a separate unit on the campus of a deaf school is suggested.

LEENHOUTS, M.A. The mentally retarded deaf children. *Proceedings: American Instructors of the Deaf*, 55-64, 1959.

The problems of the MR deaf child in California and in its residential schools are discussed.

LOWELL, E.L. A point of view regarding the multiple handicapped deaf. *Proceedings: American Instructors of the Deaf*, 40:64-69, 1961.

A discussion of the education of the multiply handicapped deaf concludes that they should not be educated in regular schools for the deaf.

LUSZKI, W.A. Application of deprivation concepts to the deaf retarded. *Mental Retardation/MR*, 2(3):164-170, 1964.

Data on effects of early deprivation and its influence on MR deaf are described in terms of mother-child relationships, cognitive and learning abilities, personality development, and social relations.

NOBER, E.H. Hearing problems associated with cerebral palsy. In: Cruickshank, W.M., ed. *Cerebral Palsy: Its Individual and Community Problems*. Revised Edition, Syracuse, New York, Syracuse University Press, 1966, Chapter 6, 277-336.

Assessment of hearing problems of CP children may be complicated by the factor of MR. A training program includes orientation, hearing aid and sound discrimination training.

SCHIEFELBUSCH, R.L., & SMITH, J.O., eds. *Research in Speech and Hearing for Mentally Retarded Children*. Lawrence, Kansas, University of Kansas, 1963, 376 p.

Papers from the 1963 Conference on Research in Speech and Hearing for Mentally Retarded Children are presented with specific emphasis on training the MR child in language skills.

Incidence

HORVORTH, I.E. The "at risk" infant. *Lancet*, 2(7521):887, 1967.

Screening techniques of children with "at risk" factors reveal a large incidence of deafness in early childhood.

LLOYD, L.L., & REID, M.J. The incidence of hearing impairment in an institutionalized mentally retarded population. *American Journal of Mental Deficiency*, 71(5):746-763, 1967.

Pure tone audiometric data are reported on 638 MR children at Parsons State Hospital and Training Center. Data are given for 8 different hearing-loss criteria and in terms of type of impairment. Results indicate that hearing loss is a major problem in the habilitation of the MR.

MCDONALD, A. *Children of Very Low Birth Weight*. London, England, William Heinemann Medical Books, 1967, 124 p.

Analysis of 1,128 English children whose birth-weight was less than 1,800 gm indicated an incidence of 1.8% of perceptive deafness of moderate or severe degree.

NUDO, L.A. Comparison by age of audiological and otological findings in a residential institution for the mentally retarded: A preliminary report. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 132-154.

An analysis of 914 MR residents to determine incidence and prevalence of major audiological and otological disturbances. Data showed that 55% had hearing loss. Fifty percent of the hearing losses were sensori-neural, 42% conductive, and 8% were mixed.

WEBB, C., KINDE, S., WEBER, B., & BEEDLE, R. Incidence of hearing loss in institutionalized mental retardates. *American Journal of Mental Deficiency*, 70(4):563-568, 1966.

Validity of hearing test procedures for the MR is discussed, and recommendations are given.

MENTALLY RETARDED DEAF

WILEY, J., & JACOBS, G. The incidence and characteristics of hearing loss in institutionalized, mildly retarded population. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*, Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 155-163.

Assessment of 130 EMR persons by pure tone and speech audiometry is cited with 80% showing normal hearing. No differences were found in functional versus organically classified Ss.

Etiology

ATHANASSIADES, T., & NICHOLSPoulos, D. Complications of varicella. *Lancet*, 2(7564): 403, 1968 (Letter)

A case history of an 8-year-old boy with acoustic nerve damage which resulted in complete deafness was reported as a complication of varicella. This disorder has not been previously associated with varicella.

MONTAGUE, A.D.W. Hemolytic disease of the fetus. In: Barnes, A.C., ed. *Intrauterine Development*. Philadelphia, Pennsylvania, Lea and Febiger, 1968, 443-466.

Erythroblastosis survivors may have MR, deafness, and abnormal EEGs.

SCHNYDER, U.W., WISSLER, H., & WENDT, G.G. Eine weitere form von atypischer erythrokeratodermie mit schwerhörigkeit und cerebraler schädigung (An additional form of atypical erythrokeratodermia with deafness and cerebral damage). *Helvetica Paediatrica Acta*, 23(3):220-230, 1968.

An EMR girl had erythrokeratodermia in combination with labyrinthine deafness, psychic and somatic retardation as well as motor problems. Chromosome karyotype was normal but vitamin-A-serum was decreased.

SIEGEL, M., FUERST, H.T., & DUGGAN, W. Rubella in mother and congenital cataracts in child: Comparative data in periods with and without epidemics from 1957-1964. *Journal of the American Medical Association*, 203(9):632-636, 1968.

Incidence and prevalence of congenital cataracts, heart disease, and bone conduction deafness in offspring of rubella and other viral epidemics were surveyed. The sample included 1,526 offspring, with 731 from virus-infected mothers and 795 from control mothers. The only definite data cited are for cataracts.

VERNON, M. Characteristics associated with post-rubella deaf children: Psychological, educational and physical. *Volta Review*, 69(3):176-185, 1967.

An analysis of 129 cases of post-rubella deaf children among 1,468 deaf indicated that their educational achievement was poor in comparison to other deaf children even when IQ level was considered. A high incidence of aphasia was found for these children.

VERNON, M. Meningitis and deafness: the problem, its physical, audiological, psychological, and educational manifestation in deaf children. *Laryngoscope*, 77(10):1856-1874, 1967.

Incidence of deafness as a result of meningitis was analyzed in 1,468 applicants to the California School for the Deaf. About 8% (114) had postmeningitic deafness with a sex ratio of 82 males to 32 females. Average CA of onset was 20 months; a critical factor since it predated speech development.

VERNON, M. Prematurity and deafness: the magnitude and nature of the problem among deaf children. *Exceptional Children*, 33(5): 289-297, 1967.

An analysis of 1,468 profoundly hard-of-hearing children was made to determine relationship between prematurity and deafness. Variables such as multiple handicaps, achievement, adjustment, IQ, audiometric responses, and brain damage were considered. Of the 25% born prematurely, 16 (3%) had IQs below 70. Premature deaf children had more serious educational problems.

VERNON, M. Rh factor and deafness: The problem, its psychological, physical, and educational manifestations. *Exceptional Children*, 34(1):5-12, 1967.

Forty-five children with deafness from erythroblastosis fetalis were surveyed for deficits in IQ, achievement, audiometry, and adjustment to determine behavioral correlates of neurological damage from erythroblastosis fetalis. The mean IQ was below the level of the general population and that of the non-Rh deaf. Over 1/4 were unable to remain in the special school for deaf children.

Assessment and Diagnosis

DANSINGER, S., & MADOW, A.A. Verbal auditory screening with the mentally retarded. *American Journal of Mental Deficiency*, 71(3):387-392, 1966.

Verbal auditory screening for children is cited as effective in detecting hearing impairment among institutionalized MR. Of 967 MR children and adults, 11% were untestable and 12% of those testable had hearing impairments which interfered with communication.

FULTON, R.T. Standard puretone and Bekesy audiometric measures with the mentally retarded. *American Journal of Mental Deficiency*, 72(1):60-73, 1967.

Data from the standard pure-tone and Bekesy audiometric procedures given to 153 institutionalized MR Ss from 4 levels of intelligence are reported. Valid results from the Bekesy procedures were obtained for lower intelligence levels.

GOETZINGER, C.P., WELLS, R.C., & DEKKER, L.C. Non-language IQ tests used with deaf pupils. *Volta Review*, 69(8):500-506, 1967.

Reliability was evaluated for 3 non-language intelligence tests employed with deaf pupils. Lower scores on the Raven Progressive Matrices Test and Terman Non-Language Multi-Mental Test were due to lack of experiences rather than to language deficit. These scales and the Chicago Non-Verbal Examination were as reliable for deaf as for hearing Ss.

HARLOW, J.L. Mentally Retarded or Hearing Impaired? *Volta Review*, 69(10):664-667, 1967.

Assessment of 3 children in a state institution for the MR is described in terms of problems of differentiation between MR and hearing impairment.

KIMMICK, H.M. Otologic aspects of mental retardation: Implications for diagnostic audiology. In: Lloyd, L.L., & Frisina, D. R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 119-136.

The status of otology in institutions for the MR is discussed. There is a lack of otologic care for the MR even though their needs are great. An audiologist can aid by screening patients for otologic services.

KOPATIC, N.J. The reliability of pure tone audiometry with the mentally retarded: Some practical and theoretical considerations. *Training School Bulletin*, 60(3):130-137, 1963.

The use of pure tone audiometry with 47 female institutionalized MRs is reported. The reliability of this technique is assured with repeated measurement. Incidence of hearing loss for MRs was considered no greater than for normals.

KRAUS, M.J., Jr. The use of psychological tests with the mentally retarded deaf. Paper presented at the 91st annual meeting of the American Association on Mental Deficiency, Denver, Colorado, May 15-20, 1967, 3 p. Mimeographed.

The validity of psychological tests for the MR deaf is discussed. The Wechsler, Raven Colored Progressive Matrices, and the Nebraska Test of Learning Aptitude are the most successful tests. However, observation of behavior for long periods of time is preferred.

LaCROSSE, E.L. A method to test the hearing of mentally retarded children. *Volta Review*, 66(1):27-31, 1964.

Instrumental conditioning procedures indicated an incidence of 15-36% of hearing loss for 384 moderately retarded children. This incidence is lower than those reported in other studies.

LAMB, N.L., & GRAHAM, J.T. GSR audiometry with mentally retarded adult males. *American Journal of Mental Deficiency*, 72(5):721-727, 1968.

Modification of galvanic skin response audiometry for MR adult males is reported in an analysis of 20 normal male adults and 20 MR male adults. GSR threshold was found for 40% of MR Ss and 85% of normals. GSR audiometry has limited use in the assessment of the MR's hearing.

LLOYD, L.L. Comparisons of selected auditory measures of mentally retarded children: A preliminary report. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 99-118.

The reliability and validity of 6 conventional audiometric techniques with MR children was assessed. Standard methods were reliable and valid. Speech audiometry appears to be more reliable than pure-tone methods.

LLOYD, L.L., & FRISINA, D.R., eds. *The Audiological Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, 314 p. Mimeographed.

Innovative techniques for the audiological assessment of "difficult to test" MR children are described. Emphasis is on operant conditioning principles.

LLOYD, L.L., SPRADLIN, J.E., & REID, M.J. An operant audiometric procedure for difficult-to-test patients. *Journal of Speech and Hearing Disorders*, 33(3):236-245, 1968.

Tangible reinforcement operant conditioning audiometry can be used to test nonverbal groups of children.

MYERSON, L. Pathways to future in the audiological assessment of the mentally retarded. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 19-31.

Proceedings of an audiological workshop held at Parsons State Hospital and Training Center are reported and problems involved in audiological assessment of the MR are discussed. Major factors were audiological phenomena in MR population, scope of assessment procedures, new techniques for hearing assessment, and testing the "difficult to test" persons.

RAPIN, I., SCAROLA, L.M., & COSTA, L.D. The Purdue Pegboard as a screening test for brain damage and mental retardation in non-verbal children. *Volta Review*, 69(10):635-639, 1967.

The validity of the Purdue Pegboard as a screening device for brain damage and MR was assessed for 74 non-verbal children. Results indicated that the test identifies some, but not all, brain damaged children and is most efficient when brain damage is associated with sensory-motor or intellectual deficits.

RENEAU, J.P., & MAST, R. Telemetric EEG audiometry instrumentation for use with the profoundly retarded. *American Journal of Mental Deficiency*, 72(4):506-511, 1968.

An EEG telemetry system can be used to assess audition and behavior in SMRs and provides an empirical approach to auditory assessment.

RITVO, E.R., ORNITZ, E.M., & WALTER, R.D. Clinical application of the auditory averaged evoked response at sleep onset in the diagnosis of deafness. *Pediatrics*, 40(6):1003-1008, 1967.

A diagnostic technique for determining hearing deficits in disturbed children employed EEGs recorded with auditory stimuli during early stages of sleep.

ROBSON, P. The "at risk" infant. *Lancet*, 2(7521):886-887, 1967. (Letter)

The "at risk" concept of infant screening was described and it was concluded 75% of CP cases, 25% of MR, and most cases of deafness could be determined 1-4 years sooner with employment of this procedure.

RYAN, M.D., & STEWART, J.H. The use of textured pictures as a reinforcement in meaningful sound identification audiometry. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 89-97.

The employment of animal sounds as stimuli and textured animal pictures as reinforcers were successful in the assessment of young MR children. Positive results appeared to be due to easier association of the stimuli (animals) and the high interest level produced by the textured animal pictures.

VITTENSON, L.K. Premature and erroneous diagnosis of mental retardation in infants with hearing and speech deficits. *Eye, Ear, Nose and Throat Monthly*, 47(9):411-414, 1968.

Diagnostic procedures for infants with multiple handicaps should be reorganized to allow for more accuracy. Many deaf children have been misdiagnosed as MR.

WALDON, E.F. Testing of infants: Implications for audio-reflexometry. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 171-191.

Employment of audioreflexometry using Baby Cry Test signals is a valid and beneficial method for determining the hearing thresholds of very young children and subnormal children.

WEAVER, R.M. The use of filmstrip stories in slide show audiometry. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 71-88.

Filmstrip audiometry is analyzed for 60 verbal and ambulatory MR Ss. This audiometric conditioning technique is a modification of the slide show conditioning technique and is recommended for testing EMR or TMR persons.

WHIPPLE, C.I. Behavior observation in the audiologic evaluation of the mentally retarded. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 193-212.

Relevant variables in the assessment of hearing in the MR may include inappropriate testing procedures, poor rapport, task difficulty, and poor socialization.

YOUNG, E., & ESTES, J. An investigation of acoustic impedance measurements in an adult mentally retarded population. Paper presented at the 91st annual meeting of the American Association on Mental Deficiency, Denver, Colorado, May 15-20, 1967, 8 p. Mimeographed.

Acoustic impedance measurements were investigated in adult MRs. The procedure is beneficial with this population because auditory functioning can be analyzed regardless of the S's ability to respond voluntarily.

Clinical Characteristics

CLAUSEN, J. Threshold for pure tone and speech in retardates. *American Journal of Mental Deficiency*, 70(4):556-562, 1966.

Pure tone thresholds and thresholds for speech were sampled for 3 groups of retardates with various age levels and 1 normal group. Pure tone thresholds appeared to be more effective than speech thresholds for hearing assessments. There was a low correlation between the 2 measures.

MENTALLY RETARDED DEAF

DOCTOR, P.V. Multiple handicaps. *Proceedings American Instructors of the Deaf*, Volume 38, 34-36, 1959.

Analysis of the educational problems of the multiply handicapped indicates a need for the development of new teaching techniques.

FLOWER, R.M., VIEHWEG, R., & RUZICKA, W.R. The communicative disorders of children with kernicteric athetosis: I. Auditory disorders. *Journal of Speech and Hearing Disorders*, 31(1):41-59, 1966.

Analysis of communication disorders of 15 children with kernicteric athetosis and reduced auditory sensitivity was made. Data are reported on audiograms, auditory discrimination tests, and auditory visual recall.

HODGSON, W.R. Auditory characteristics of post-rubella impairment. *Volta Review*, 71(2): 97-103, 1969.

Case histories illustrate that post-rubella hearing-impaired children appear to benefit from wearing hearing aids and can tolerate amplified sound.

HODGSON, W.R. Audiological report of a patient with left hemispherectomy. *Journal of Speech and Hearing Disorders*, 32(1):39-45, 1967.

A hemispherectomy resulted in no significant differences in the motor, speech, or hearing behavior of a 17-year-old MR girl with right infantile hemiparesis.

KEIR, E.H. Communication problems of the mentally retarded deaf child. In: Van Pelt, J.D., ed. *Proceedings of the Fourth Interstate Conference on Mental Deficiency*. Melbourne, Australia, Australian Group for the Scientific Study of Mental Deficiency, 1965, p. 43-50.

Characteristics of 12 young MR children in Melbourne (Australia) were presented. The etiologies of 10 of the 12 indicated a risk for deafness. Speech is seldom acquired by the MR deaf child. Early meaningful relationships are the most important prerequisite for communication.

LAGUAITE, J.K., & JOSEPH, M. A study of children with communication problems associated with maternal rubella. *Southern Medical Journal*, 58(2):231-235, 1965.

Observations of 39 hearing impaired children were reported for a 9-year-period. Data indicated that 28 mothers had rubella during pregnancy. Twenty-three of these children could have profited from early training for the deaf.

LESHIN, G.J., & STAHLCKER, L.V. Academic expectancies of slow learning deaf. *Volta Review*, 64(10):599-603, 1962.

A discussion of academic expectancies for slow learning deaf children is presented with emphasis on their poor achievement level. The IQ range for MR is defined as 0 to 90.

LUSZKI, W. Hearing loss and intelligence among retardates. *American Journal of Mental Deficiency*, 70(1):93-101, 1965.

The relationship between scores on the WAIS and WISC and hearing loss of retardates from the Michigan Deaf Retarded Project was described. Results showed no differences among Ss for 5 hearing levels ranging from normal to no hearing except for the Block Design Subtest.

NELSON, M., & SIBILIO, J.P. Audiological aspects of a deaf retarded population. *Volta Review*, 64(7):426-427, 1962.

Audiological problems of the MR deaf patient in the State Home and Training Schools of Michigan are summarized.

PRINGLE, M.L.K. The emotional and social needs of deaf and brain-damaged children. *Teacher of the Deaf*, 63:13-21, 1965.

Favorable social and emotional adjustment of deaf and brain-damaged children are related to the attitudes and insight of parents and teachers. These children need love, security, new experiences, and recognition, achievement, and responsibility.

ROOF, R.L., LONGMORE, J.B., & FORRESTER, R.M. A childhood syndrome of bone dysplasia, retinal detachment and deafness. *Developmental Medicine and Child Neurology*, 9(4):464-473, 1967.

A syndrome was described in case reports of 4 children with bone dysplasia, dwarfism, pseudoglioma, blindness, and deafness. Hearing defects were variable. The one child who evidenced MR appeared more dwarfed and less deaf and may not truly represent this syndrome.

SIMPSON, P.E. The problems of the backward deaf child. *Teacher of the Deaf*, 59:232-236, 1961.

A group of 150-300 British deaf children were surveyed to determine reasons for lack of academic progress.

VAN PELT, J.D. *Proceedings of the Fourth Interstate Conference on Mental Deficiency*. Melbourne, Australia, Australian Group for the Scientific Study of Mental Deficiency, 1965, 100 p.

Communication problems of the MR deaf child are discussed in the program implemented by the Australian Group for the Scientific Study of Mental Deficiency. This program was established to aid inter-disciplinary communication in the field of MR in Australia.

WARREN, S.A., & KRAUS, M.J., JR. Deaf children, mental retardation and academic expectancies. *Volta Review*, 65(7):351-352, 1963.

Critical comments on an article by Leshin and Stahlecker are directed toward their definition of MR as well as their conclusions that the retarded deaf child should be educated in an institution for the MR.

WOODFORD, D.E. An investigation into problems of the deaf and partially deaf children with additional handicaps. *Teacher of the Deaf*, 60:120-128, 1962.

A report for the National College of Teachers of the Deaf in England investigates the problems of the multiply-handicapped deaf.

Education and Therapy

ANDERSON, R.M., & STEVENS, G.D. Qualifications of teachers of mentally retarded deaf pupils in residential schools for the deaf. *Special Education in Canada*, 43(2):23-32, 1969.

A survey of 150 teachers from 64 residential schools for the deaf indicated that expediency rather than training determined assignments as teachers of MR deaf pupils. Forty-three percent of these teachers had a hearing loss. Most felt inadequately prepared for the problems of this population, and 66% preferred to teach deaf children with no other disability.

ANDERSON, ROBERT M., & STEVENS, GODFREY D. Policies and procedures for admission of mentally retarded deaf children to residential schools for the deaf. *American Annals of the Deaf*, 115(1):30-36, 1970.

Data from 64 residential schools for the deaf show that policies for admission of MR deaf children are variable and unclear. The local administrators determine the criteria, and often they prefer not to admit children with more than one handicap. Re-examination of admission criteria is suggested.

BASIL, R.A. Hearing conservation aspects of mental retardation. In: Lloyd, L.L., & Frisina, D.R., eds. *The Audiologic Assessment of the Mentally Retarded: Proceedings of a National Conference*. Parsons, Kansas, Parsons State Hospital and Training Center, Speech and Hearing Department, 1965, p. 165-169.

The Michigan Hearing Conservation Program is reported. Speech and hearing services for the MR in Michigan have been expanded.

CANDLAND, D.K., & CONKLYN, D.H. Use of the oddity problem in teaching mentally retarded deaf mutes to read: A pilot project. *Training School Bulletin*, 59(2):38-41, 1962.

In teaching MR deaf mutes to read, the "oddity problem" technique was successful.

MENTALLY RETARDED DEAF

CHAMBERLAIN, N.H. A HOPE special education program in Guinea. *Exceptional Children*, 33(3):177-178, 1966.

Special education programs initiated in Conakry, Guinea, West Africa, by Project HOPE are described and include classes for MRs with hearing losses.

GLOVSKY, L., & RIGRODSKY, S. A classroom program for auditorially handicapped mentally deficient children. *Training School Bulletin*, 60(2):56-69, 1963.

A special class with a communication-oriented program for auditorially impaired MR children at Vineland Training School is described in terms of Ss and techniques.

GREENBERG, S.I. Speech and language therapy with hearing impaired multiply handicapped mentally retarded children. *Welfare Reporter*, 19(1):20-24, 1968.

Training for hearing impaired, multiply handicapped MR children at the North Jersey Training School progresses from gross sound stimulation thru gross sound identification, discrimination, and localization to patterning of sound stimuli and individual therapy. Successful methods are described.

HENRIELLA, M. The slow learning deaf child: Part I. *Volta Review*, 63(8):380-384, 1961.

Methods of instruction for the MR deaf child with emphasis on drill, teacher organization, and planning are presented.

HENRIELLA, M. The slow learning deaf child: Part II. *Volta Review*, 63(9):444-448, 1961.

Assessment problems related to the slow learning deaf child are described. MA rather than IQ should be primary criteria.

HURWITZ, S.N., & DeFRANCISCO, S. Behavioral modification of the emotionally retarded deaf. *Rehabilitation Literature*, 29(9):258-264, 1968.

A 3-year rehabilitation program for 165 deaf clients is described. A socio-behavioral

approach involving environmental structuring is preferred. A relevant, realistic, and consistent approach with introduction of positive reinforcement is suggested. Results show that 2 of 3 clients have found employment.

McCOY, D.F., & LLOYD, L.L. A hearing aid orientation program for mentally retarded children. *Training School Bulletin*, 64(1):21-30, 1967.

A clinical hearing aid orientation program for EMR and TMR institutionalized children with mild to severe hearing impairments is described. Cottage personnel are instructed in how to teach the child to use and care for the hearing instrument.

MITRA, SUDHANSU B. Educational provisions for mentally retarded deaf students in residential institutions for the retarded. *Volta Review*, 72(4):225-236, 1970.

A survey of facilities for the MR deaf children in state residential facilities for the retarded showed that their diagnostic techniques varied and 50% of the reporting teachers had no training in working with the deaf. Major problems involve identification, evaluation, and audiological testing of MR deaf and teacher preparation for this specific population. Smooth transition of students between institutions for the deaf and those for the MR should be available.

PENNSYLVANIA SCHOOL FOR THE DEAF. *Curriculum Guide Lines for the Slow-Learning Deaf Child at the Pennsylvania School for the Deaf*. Philadelphia, Pennsylvania, 1965, 61 p.

A curriculum which emphasized language and speech activities was developed for deaf slow learners. An appropriate vocabulary check list is needed to record the child's progress in language development.

SCHLANGER, B.B. *The Effects of Listening Training On The Auditory Thresholds of Mentally Retarded Children*. Morgantown, West Virginia, West Virginia University, 1961, 116 p.

Incidence of hearing loss in MR children and the effects of listening training were studied.

SELLIN, D.F. The mentally retarded, hearing handicapped learner: Implications for teacher education. *Volta Review*, 66:258-261, 1964.

Problems related to educational programs for MR deaf children in residential schools are overlooked by teachers. The program of one residential school is described as effective.

SILVERMAN, S.I. Speech and language therapy with hearing impaired, multiple handicapped mentally retarded children. Paper presented at the 91st annual meeting of the American Association on Mental Deficiency, Denver, Colorado, May 15-20, 1967, 12 p.

Therapeutic goal of treatment for hearing impaired multiply handicapped MR girls is to stimulate and develop language skills to improve communication. Specific goals include awareness, meaning, and employment of auditory stimuli.

STOLP, L.E. A curriculum for the slow-learning deaf child. Available from the Alexander Graham Bell Association for the Deaf, Reprint Number 824, 1965, 5 p.

A curriculum for slow-learning deaf children is discussed with suggestions for self contained units of 7-8 students for each teacher. A curriculum should be dynamic and based on students' needs.

STUCKLESS, E.R., & BURROWS, N.L. Teaching methods with the mentally retarded deaf student. Paper presented at the 91st annual meeting of the American Association on Mental Deficiency. Denver, Colorado, May 15-20, 1967, 26 p. (Mimeographed).

Diagnosis, classification, therapy, and curriculum are variables to consider when developing teaching methods for the MR deaf. Borderline MRs with moderate hearing losses may achieve at the EMR level and can be taught in special classes.

TAYLOR, A.P., & POLLOCK, B.E. A structured program of learning for moderately retarded deaf adults. *Volta Review*, 70(2):114-117, 1968.

A structured program designed to improve the communication of 2 institutionalized women with IQs of 83 and 61 is reported. A speech therapist and a teacher were employed for 16 hours a week, for 8 weeks. The program focused on time concepts, language arts, and social training. Results were positive.

UNITED STATES. HEALTH, EDUCATION and WELFARE DEPARTMENT. *A Review of Selected Program Activities in the Education of the Deaf*. Washington, D.C., Superintendent of Documents, U.S. Government Printing Office, 1967, 18 p.

The allocation of funds under federal programs designed for MR, hard of hearing, deaf, speech impaired, and multi-handicapped children are discussed.

Addendum

Additional references without annotations

ABRUZZO, A., et al. The Identification and Vocational Training of the Institutionalized Retarded Deaf Patient. Office of Vocational Rehabilitation, RD 800, Washington, D.C., 1961.

JAMES, W.C. Mentally retarded deaf children in a California state hospital. In: *Report of the Proceedings of the Forty-first Meeting of the Convention of American Instructors of the Deaf*, Gallaudet College, Washington, D. C. June 1963. Washington D.C., Superintendent of Documents, U.S. Government Printing Office, 1964, p. 573-577.

JOHNSON, R. K. The institutionalized mentally retarded deaf. In: *Report of the Proceedings of the Forty-first Meeting of the Convention of American Instructors of the Deaf*, Gallaudet College, Washington D.C., June 1963. Washington, D.C., Superintendent of Documents. U.S. Government Printing Office, 1964, p. 568-573.

LEENHOUTS, M. A. The mentally retarded deaf child. In: *Report of the Proceedings of the Thirty-ninth Meeting of the Convention of American Instructors of the Deaf*. Colorado Springs, Colorado, June, 1959. Washington, D.C., Superintendent of Documents, U.S. Government Printing Office, 1960, 55 p.

MANGAN, K. R. A state program of services for the mentally retarded deaf child. In: *Report of the Proceedings of the Forty-first Meeting of the Convention of American Instructors of the Deaf*, Gallaudet College, Washington, D. C., June 1963. Washington D. C. Superintendent of Documents, U. S. Government Printing Office, 1964, p. 565-568.

MENTALLY RETARDED DEAF

WOLFE, W. G., & HARVEY, J. E. A Comparative Investigation of Methods of Testing Auditory and Visual Acuity of Trainable Mentally Retarded Children. Part II. Washington, D.C., United States Office of Education, Cooperative Research Branch, SAE-6477-1960.

WOLFE, W. G., & MacPHERSON, J. R. A Comparative Investigation of Methods of Testing Auditory and Visual Acuity of Trainable Mentally Retarded Children, Part I. Washington, D.C., United States Office of Education, Cooperative Research Branch, SAE-6477-1960.

BROAD ASPECTS OF MENTAL RETARDATION

- 2133 DALY, WILLIAM C., & BELLAMY, EDWARD E.
Historic milestones in mental retardation: Random notes and sketches. *Clinical Pediatrics*, 8(9):543-547, 1969.

Prior to the middle ages, historic documents carrying references to what is now called MR were rare; however, most handicaps were viewed as hopeless and the defective young were destroyed or abandoned. During the middle ages, court personages were entertained by jesters and buffoons, many of whom were MR. The Protestant Reformation ushered in a different set of attitudes in which it was thought that retardates were possessed by devils. Around 1800, Jean Itard began an intensive program to civilize a wild boy who had been diagnosed as MR. Although Itard considered himself to have failed in his efforts, he aroused new concern for the MR in professionals. Edouard Seguin continued Itard's work and was frequently consulted regarding the opening of residential schools. The period from 1850 to 1900 is often referred to as the "institutional period" and was characterized by construction of large residential institutions. During 1900, special classes for the MR were begun. Between 1910 and 1935, these special classes expanded, institutions continued to increase in size, and programs that originated years before were implemented. In 1950, the National Association for Retarded Children was established. Recently, laws have been passed which favor MR educational programs, decentralization of residential schools, maximized individual treatment, and establishment of day care centers, workshops, camps, and activity centers. In 1961, President Kennedy organized a panel to study the problem of MR on a national level, and 1963 saw the beginning of the allocation of vast federal funds to explore possible solutions and to research all areas of MR. (5 refs.) - B. Parker.

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- 2134 CARDNO, J. A. Idiocy, imbecility: An early American contrast. *Psychological Record*, 18(2):241-245, 1968.

The definition and description of low level retardation by Isaac Ray, an institution superintendent, and T. S. Upham, a philosopher, are compared. Both were Americans who wrote in the mid-nineteenth century on the topic of retardation. Ray made a greater attempt to distinguish classes within the MR population, and he provided more in-depth descriptions; however, he stopped short of suggestions for testing or measurement. Upham, on the other hand, approached the subject in less precise terms and was more philosophical. (8 refs.) - J. M. Gardner.

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Hobart, Australia

- 2135 SECRETARY'S COMMITTEE ON MENTAL RETARDATION. *The Problem of Mental Retardation*. Washington, D. C., Superintendent of Documents, Government Printing Office, 1969, 18 p. \$0.20.

The problem of mental retardation in the United States is outlined with information given on the incidence, definition, and levels of retardation. A discussion of the difference between mental retardation and mental illness is also presented. A brief discussion is given of community services, manpower, residential care, special education, rehabilitation, and preventive measures. (No refs.) J. M. Gardner.

- 2136 PRESIDENT'S COMMITTEE ON MENTAL RETARDATION. *MR 69: Toward Progress: The Story of a Decade*. Washington, D. C., Superintendent of Documents, Government Printing Office, 1969, 32 p. \$0.60.

Progress in mental retardation treatment, services, and research is summarized, including information on increased assistance to poverty areas, improved manpower recruitment and training programs, and public-private partnerships in planning. Recommendations for local, state, and national action are made in the areas of habilitation and employment, poverty, education, and research. A declaration of general and special rights of the MR is included. (No refs.) - J. M. Gardner.

- 2137 JERVIS, GEORGE A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, 262 p. \$12.50.

This collection of papers from the Third Scientific Symposium of the Joseph P. Kennedy Jr. Foundation held in 1966 includes data on the genetics of MR; genetic, chemical, and anatomical factors in the development of the nervous system; the etiological effects of environmental factors; the nature of learning; problems of rehabilitation; and the physical performance of MRs. Environmental factors discussed include undernutrition and early intellectual deprivation. The results of early educational intervention programs are presented. Discussions of the biological bases of learning and modern techniques of learning which emphasize operant techniques are included. Papers on rehabilitation in MR deal with socioeconomic factors, the effects of institutionalization, community adjustment, a community program, and the rehabilitation and employment of SMRs. Papers on physical performance and recreation include data on EMRs, TMRs, and SMRs as well as on diagnosis and prescription. This book would be of interest to geneticists, biochemists, neurologists, nutrition specialists, physical educators, special educators, biologists, psychologists, hearing specialists, recreation specialists, physicians, and social workers. (358 refs.) J. K. Wyatt.

CONTENTS: Genetics of Mental Retardation; Problems of Neurogenesis; Nutrition and the Development of Nervous Systems; Deprivation Factors in Mental Retardation; Biological Basis of Learning; Operant Techniques in Mental Retardation; Rehabilitation in Mental Retardation; Physical Performances of the Mentally Retarded.

- 2138 LEMKAU, PAUL V., & IMRE, PAUL D. Results of a field epidemiologic study. *American Journal of Mental Deficiency*, 73(6):858-863, 1969.

Previous studies on the incidence of MR reported varying figures which range from 0.4 to 9.0/100 population. This was due to different data collection techniques and to the use of outdated census figures. A small (under 20,000) Southeastern United States county where current census figures were available was selected to study. Census forms were mailed to each household, and trained interviewers went to each home, picked up and edited the census and completed a Vineland-like behavior check list on all children over one year of age. Of 2,266 behavior checklist profiles, 592 were further screened, and 116 (or 5.3%) had IQs less than 69. Examination of school children with IQs less than 79 (based on a group IQ test) with the Stanford-Binet revealed the following MR rates for varying ages: 6.6% (CA 6 to 9); 8.9% (CA 10 to 14); and 9.6% (CA 15 to 19). The peak incidence was in the oldest group and not in the 10 to 14 year range which is the usual finding. Of the 7,382 children aged 1 to 19 screened, 549 (or 7.4%) had IQs less than 69. An attempt to obtain follow-up data is being made. (12 refs.) - J. M. Gardner.

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- 2139 CONACHER, D. G. Mental retardation in Angus County, Scotland. *American Journal of Mental Deficiency*, 73(6):970-980, 1969.

In a survey of 205 MRs who reported to the Strathmartine Hospital in Angus County, Scotland in 1967, it was found that severely retarded Ss (IQ < 50) were evenly distributed across socioeconomic ranges; however, mildly retarded Ss were proportionately greater in the semi-skilled and unskilled groups. The rate of retardation was greatest in the 15-19 year age range. The greatest percentage of patients were in the moderately retarded range (41%). Almost 50% of the patients had unknown etiologies. This was less true of persons with IQs less than 50 (33%) as compared to those over 50 (76%). Prevalence rates/10,000 for various conditions were: Down's syndrome 42, cerebral palsy 26, epilepsy 13, and PKU 3. The major cause of institutionalization among mildly retarded persons was inappropriate social behavior (38%). (25 refs.) J. M. Gardner.

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- 2140 JOSE, D. G., SELF, M. H. R., & STALLMAN, N. D. A survey of children and adolescents on Queensland aboriginal settlements, 1967. *Australian Paediatric Journal*, 5(2): 71-88, 1969.

A survey of 12 aboriginal communities in Queensland indicates that mortality and morbidity rates are 6 times greater than the Queensland average, particularly in the pre-school age group. The survey was made from May to November, 1967, and included 12 settlements and missions with a population of 8,000. More than 1/2 of the Ss were less than 20 years old. Records were available for 11 of the 12 settlements for the 5 years from 1962 through 1966. The reported cause of death of aboriginal infants and children on 8 settlements during 1962-1966 included congenital malformation, encephalitis, cerebral abscess, meningitis, prematurity, malnutrition, and status epilepticus. Growth curves of settlement children indicated growth retardation after 6 months of age. Infections of the respiratory tract, urinary tract, and skin were also common. There was a high proportion of rheumatic heart disease and non-organic heart murmurs. Hepatomegaly was found in 11.3% of the children. Congenital malformations included conditions with MR, CP, hydrocephalus, and cataracts. Erythrocyte sedimentation rates were high in more than 1/2 the children tested. Hemoglobin values were lower than in Australian normals. Cold agglutinins were found in 20% of the sample in low titers and in about 1/2 the children tested in 2 settlements. Only 1% of the children had positive evidence of treponemal infection. (68 refs.) - F. J. McNulty.

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- 2141 CRANDELL, JOHN M., JR. The genesis and modification of attitudes toward the child who is different. *Training School Bulletin*, 66(2):72-79, 1969.

Attitudes are defined as a special class of concepts in which affective components predominate. Concepts are viewed as mental structures in which affective, cognitive, and behavioral components are present in varying amounts. It is postulated that affective components arise from perceptions of environmental perceptions which are ambiguous, incongruous, or strange. Exceptional children (those with physical or behavior deviations) are cited as examples of affects arousal. Although one's reactions to these elements may be changed through learning, they are fundamentally reflexive and are more difficult to

change than are cognitive or behavioral elements. Evidence from research in the behavioral science is presented which tends to support the theory. Suggestions are made for modifying attitudes through either varying the levels of cognitive and behavioral components, or through providing affective experiences which will change negative affect to positive. (16 refs.) - *Journal abstract*.

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- 2142 ORZACK, LOUIS H. Social changes, minorities, and the mentally retarded. *Mental Retardation/MR*, 7(5):2-6, 1969.

Social attitudes toward the retarded and opportunities provided them have fluctuated widely. Achievement, impersonality, and the growth of organizations create difficulties for the handicapped. Optimism about potentials and discriminatory treatment of the retarded and of other minorities have co-existed in our culture. Increased support for change comes from special groups, spokesmen for the retarded, but obstacles and discriminatory barriers that help generate retardation continue to exist. (10 refs.) - *Journal abstract*.

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- 2143 Hope for the mentally retarded. *Royal Bank of Canada Monthly Letter*, 50(10): 1-4, 1969.

Canadians believe that a majority of MRs can be successfully integrated into society. In Canada, each child is entitled to every opportunity to enhance his growth and development. Research is directed toward prevention, treatment, and care of the MR, and the Canadian government recognizes that mentally handicapped children are entitled to the same privileges, opportunities, and protection as other citizens. Parents, schools, churches, professionals, and the Government are acknowledging their responsibility to provide the environment in which MR children can become useful and productive citizens. Early detection and the guidance of professionals enables parents to help their child have equal opportunities within limitations and to engage in total, structured, and organized planning. Adequate programming must be made to insure continuous care and supervision of

the MR throughout his lifetime and should include provisions for guardianship upon the death of the parents. The Canadian Association for the MR has played a significant role in the establishment of diagnostic centers, clinics, treatment, training, and educational facilities. It has a program of demonstration projects covering every province, and the Kinsmen National Institute on MR will be a clearinghouse for all research findings in Canada and abroad. Local associations working at the community level have succeeded in gaining increased acceptance of the MR in their communities, have provided volunteers, and have encouraged fund-raising drives. (No refs.) - S. Half.

- 2144 WHO expert committee on medical rehabilitation: Second report. (World Health Organization Technical Report Series, Number 419). Geneva, Switzerland, World Health Organization, 1969, 23 p. \$0.60.

The World Health Organization Expert Committee on Medical Rehabilitation met in 1968 to define and make recommendations on medical, social, and vocational rehabilitation of the disabled. Countries should pass legislation to establish and finance services before gathering statistics on types and number of disabilities. Services should be started in hospitals, and specialists should be available to aid hospital personnel to treat in- and outpatients. The rehabilitation staff should have physicians, nurses, therapists, psychologists, counselors, special educators, placement personnel, prosthetists, and orthotists. Physicians and nurses should be trained in rehabilitation medicine as soon as possible. Schools are needed to train therapists; however, in-service training can be an immediate answer. Research centers should be set up with pilot rehabilitation centers and in conjunction with universities. (12 refs.) M. Flessinger.

- 2145 WASHINGTON. INSTITUTIONS DEPARTMENT, & UNIVERSITY OF WASHINGTON. *Proceedings of the Ninth Annual Research Meeting. Olympia Washington, Research Report*, 2(2):1-160, 1969.

The papers included in this report cover a wide range of research and recommendations in the areas of medicine, education, special education, psychology, mental health, rehabilitation, nursing, sociology, and social work. Reports on the MR include a description of diagnostic services provided by Child Study Clinics (Washington), a discussion of the

life cycle of families of MR children, research findings on the effects of behavior modification techniques in several areas of development with several different populations, and an academic curriculum for SMRs. (165 refs.) - J. K. Wyatt.

- 2146 SCHWERDT, JOHN. Architecture and mental subnormality: IV. Therapeutic variety--A day-to-day basis of design for the subnormal. *Journal of Mental Subnormality*, 14(2):101-103, 1968.

The architect can provide therapeutic variety in buildings used in training MRs by providing a physical environment which prepares the MR for situations found outside the educational milieu. The teaching situation should extend beyond the central tasks and include all the student's living, working, and learning. Functional variety can be used to stimulate and protect the MR. Space around the building can be used for educative purpose. Some experience may have to be imported if the center is in a rural area. In recognizing that the MR reacts more slowly to an experience than does the normal and becomes puzzled more readily, the architect can specifically plan variety in the building and in situations in an effort to prepare the MR for setbacks when he meets these experiences elsewhere. Easily amassed feedback analysis can be applied to further coordinating efforts in developing therapeutic architecture. (2 refs.) G. M. Niern.

No address

- 2147 BLAND, G. A. Architecture and mental subnormality: VI--Some architectural requirements for a scientific approach to teaching the mentally handicapped in hospital schools. *Journal of Mental Subnormality*, 15(2):103-109, 1969.

Hospital schools must be designed around special curricula and environments which are needed to stimulate the MR. Current hospital schools cause social and mental regression because conventional classroom and teacher philosophies are outdated. A study of MRs (CA 4-16 yrs) attending school in 34 hospitals showed: 43.4% had IQs below 30; 46.8% had IQs between 30 and 55; 48.3% had speech defects; 16.5% had gross motor defects; and 9.4% had defects of hearing or vision. The nurse:patient ratio varied from 1:3 to 1:50; one hospital had paints, and 2 had books on the ward. Most schools lacked rooms for music, home economics, or art and crafts nor did they

have libraries, gardens, or swimming facilities. The architect must provide for a stimulating, adaptable atmosphere which augments the teacher's new role in: the diagnosis of MR needs; observation; programing for each MR; evaluation of progress; and stimulation of exploration by the MR. Classrooms should be designed for 8 to 10 MRs with special furniture, carpeted areas, private corners, and easy access to an outdoor play area. Other rooms should contain accessible paints, craft materials, clay, swimming pool, home economics unit, and a gym. All surfaces should be varied and outdoors areas need different plants and land contours. Special areas are needed for SMRs who need protective care. In addition, an area with quiet rooms for individual instruction and teaching machines should be provided as well as: a senior section with more privacy and workshop activity rooms; a home economics unit; a staffroom with records, duplicating machines, and seminar space; a clinical room for testing; and a principal's office. (11 refs.) - M. Plessinger.

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- 2148 GUTMAN, ERNEST M., & GUTMAN, CAROLYN R.
Wheelchair to Independence: Architectural Barriers Eliminated. Springfield, Illinois, Charles C. Thomas, 1968, 136 p. \$6.75.

The suggestions and recommendations included in this book should enable architects and builders to adapt and alter existing residential and public structures to remove architectural barriers and to design new buildings for the benefit of the disabled. Almost 20% of the population of the United States is disabled, and by 1980, there will be one disabled person for every able-bodied person. The removal of architectural barriers will reduce the confinement of the disabled, permit them to work, and give them an opportunity for financial independence. The primary emphasis of this book is on architectural obstructions which have adverse effects on wheelchair users. There are 41 illustrations of architectural innovations. This book should be of interest to architects, builders, educators, city planners, hospital planners, and industries concerned with the employment of the handicapped. (31-item bibliog.)
J. K. Wyatt.

CONTENTS: Home is More Than Just a Wheelchair; It's Made for You; Architects and Builders, Thanks--Off Limits No Longer; Plan Wisely for Higher Learning; Down Barriers--Up Education; A Break for Disabled Small-Fry; Hospitals, Cure Thyselves! Your Telephone--A Legacy of Responsibility.

- 2149 CURRAN, WILLIAM J., STEARNS, BARBARA, & KAPLAN, HONORA. Privacy, confidentiality and other legal considerations in the establishment of a centralized health-data system. *New England Journal of Medicine*, 281(5): 241-248, 1969.

A major issue confronting health-data systems is the protection of privacy and confidentiality. State laws covering health-information disclosure are geared to specific information rather than to comprehensive coverage of all health data. Federal laws cover only data gathered by federal agencies. Every health-data system should adopt a code of ethics and clearly defined rules and regulations governing protection of information. Violation by employees should be grounds for dismissal, and violation by users should bar future access to the system. Criminal as well as civil penalties for wrongful disclosure should be adopted by statute. The system should make use of interagency agreements. A privacy committee and a method of public surveillance should be established. The organizational structure should take the form of a consortium of the contributing agencies to ensure the continuing cooperation and support of these agencies and the greatest public acceptance of the system. (20 refs.) - *Journal abstract.*

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- 2150 The Royal Medico-Psychological Association's memorandum on the Green Paper on the administrative structure of the medical services in England and Wales. *British Journal of Psychiatry*, 115(522):601-603, 1969.

Objectives for the medical services are needed before the scope of new authorities and the aspects of their organization can be determined. The administrative structure must be unified with a single authority in each area to coordinate health services. Each area board should try to guarantee adequate graduate education, public health facilities, and voluntary services. These boards should be responsible only to the Minister and the senior staff should handle day-to-day coordination. Complaints should be investigated by the Privy Council of Health Commissioners. (No refs.) - V. G. Votano.

2151 FEINGOLD, MURRAY, FAIN, TOBY, & GELLIS, SYDNEY S. Birth defect information center. *New England Journal of Medicine*, 280(16):902, 1969. (Letter)

An evaluation of the Birth Defect Information Center (Boston) after the first 400 inquiries showed that, of 260 persons answering a survey questionnaire, over 85% used the information and would call again if the need arose. Parents made up the largest category of callers (51%) and physicians almost the smallest (5%). Such a center in each state should be of great help in locating the facility with services best suited to fill the needs of each handicapped person. (No refs.) - E. F. MacGregor.

New England Medical Center Hospitals
Center for Genetic Counseling and
Birth Defects Evaluation
Boston, Massachusetts 02111

2152 LIPTON, MAY. The problem of birth defects today. *Journal of School Health*, 39(1):40-42, 1969.

Birth defects affect more than 250,000 newborn babies each year and are responsible for the death of an additional 500,000 babies before birth. While the exact causes of birth defects are unknown, it is estimated that 20% are due to heredity, 20% to environment, and the remaining 60% to some combination of heredity and environment. Environmental causes include virus infections, drugs, and X-rays. Hereditary disorders include PKU, Down's syndrome, and sickle cell anemia. Promising research is being done in the areas of Rh disease, rubella (German measles), and Hurler's disease (gargoylism). (No refs.) - J. M. Gardner.

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School Relations
National Foundation-March of Dimes
New York, New York

2153 LEWIS, MELVIN, McCOLLUM, AUDREY T., SCHWARTZ, A. HERBERT, & GRUNT, JEROME A. Informed consent in pediatric research. *Children*, 14(4):143-148, 1969.

The nature of "informed consent" and relevant issues concerning the involvement of a child in pediatric research are discussed. Nine safeguards recommended are: a review committee should assess the research design as well

as possible risks to the child; a child psychologist should participate; the investigator should have a series of personal interviews with the child in order to build trust; a social worker and a psychiatrist should be available to prepare both the child and the parents for the research; the child's care in the hospital should be planned in advance; the research team should include a pediatrician, nurse, child psychologist, and child psychiatrist or social worker to provide continuing evaluation of the child's psychological reaction to the research; the activities of the staff should be coordinated at regular interdisciplinary meetings; and follow-up care should be provided and the staff should show great discretion in releasing information on certain psychological facts gleaned from the research. If these procedures are closely followed the term "informed consent" will be more meaningful and possible risks to the child will be greatly minimized, thereby benefiting important pediatric research. (8 refs.) - B. Parker.

No address

2154 SIMS, E. B. To keep alive. *Australian Paediatric Journal*, 4(3):193-194, 1969. (Letter)

The physician and the father of a defective newborn with multiple handicaps may be justified in withholding life-saving surgery. Pledged to the saving of lives, the physician is faced with a dilemma: he may withhold removal of a duodenal obstruction in a neonate mongoloid and allow it to die. Nevertheless, there is a chance the obstruction will not cause death and the newborn may survive in an even more damaged state. The father, rather than the recently confined mother, should share the decision with the physician. The Catholic Church dictates that extraordinary means do not have to be employed to save severely deformed newborns--a precept that allows the father to decide to withhold treatment. (1 ref.) - S. Markworth.

No address

2155 JOHNSTON, STANLEY. Two kinds of tolerance. *Australian Paediatric Journal*, 4(3):194-195, 1969. (Letter)

Law and morality dictate that deformed newborns should be given life-saving measures. Legal codes make it a crime to abandon a deformed infant. Except in urgent cases, the physician must abide by parents' decisions

about treatment. In cases of inevitable perinatal morbidity, treatment should make the infant as comfortable as possible while he is alive. In cases of incurable perinatal morbidity, judgment should be made in the light of the patient's life, not the parents'. The kind of life these children will have will depend on the quality of care and rehabilitation that is available to them. (3 refs.)
S. Markworth.

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- 2156 LYNCH, HENRY T. *International Directory of Genetic Services*. Second Edition. (Birth Defects Genetic Services.) New York, New York, National Foundation-March of Dimes, 1969, 40 p.

The rapid growth of medical genetics is reflected in the current listing of genetic units. Over 560 units are listed under their respective states and countries, and the type of genetic service available is indicated for each. On a world-wide basis, 427 units (181 in the United States) offer cytogenetic service--the most frequent service offered. Clinical genetics (344 units) and genetic counseling (333 units) are the second and third most frequently offered services, while psychiatric genetics (48 units) and radiation genetics (48 units) are the least frequently offered services. (No refs.) - A. Huffer.

- 2157 *Third Annual Directory of Facilities for the Learning Disabled: 1970*. San Rafael, California, Academic Therapy Publications, 1970, 40 p.

Because the need for an up-to-date directory of facilities for learning disabled children continues, a third edition of the *Academic Therapy* directory was prepared. The facilities are listed under divisions including: schools and clinics (by state, alphabetically); summer camps (by state, alphabetically); and parent and professional organizations (alphabetically). Each facility has listed the full name, the address, and the person to contact about supplementary information. In addition, some of the schools and camps have listed the type of treatment available, age accepted, and other pertinent information. (No refs.) - M. D. Nutt.

- 2158 *Resources for the Retarded in New Jersey*. New Brunswick, New Jersey, New Jersey Association for Retarded Children, 1967, 141 p. \$2.00.

The New Jersey Association for Retarded Children has prepared its fifth edition of a comprehensive directory of resources and services for the MR. The directory encompasses extensive history, background information, interpretation of available services, eligibility requirements, source of referrals, staff, functions, purposes and total care, supervision, treatment, and management of the MR. New concepts, extended services, and opportunities are outlined and include seizure clinics, diagnostic and evaluation centers, state and private residential placements, day care units, preschool areas, adult activity centers, counseling agencies, guardianship plans, educational and vocational programs, recreational and scouting and camping facilities. It also covers the scope of rehabilitation and sheltered workshop entities. An appendix categorizes services rendered in specific counties, and a chapter is devoted to questions and answers regarding direct services available to the MR. Envisaged programs for the future are discussed. (No refs.) - S. Half.

CONTENTS: Facts About Mental Retardation; Organization of Services to the Retarded in New Jersey; Diagnostic and Evaluation Facilities and Treatment Resources; Education for the Mentally Retarded; Day Care for Children and Adults; Vocational Rehabilitation and Employment of the Retarded; Recreation and Group Activities; Residential Care; Counseling, Social Services, Guardianship and Income Maintenance; Directory of Services.

- 2159 TYMCHUK, ALEXANDER J., & KNIGHTS, ROBERT M. *A Two Thousand Item Bibliography: The Description, Etiology, Diagnosis, and Treatment of Children with Learning Disabilities or Brain Damage*. Ottawa, Canada, Carleton University, 1969, 186 p. \$3.00. (Copies available from Robert M. Knights, Carleton University)

The items in this bibliography, which included many documents concerned with MR, are arranged according to content under the following categories: bibliographies and abstracts; general reviews; general description (learning disability, minimal cerebral dysfunction, aphasic, dyslexia, hyperactivity, and cerebral palsy); etiology (neurological and educational); diagnosis (EEG, psychological, and educational); treatment (pharmacological and educational); and effects of treatment and follow-up studies. (2,064 refs.) - A. Huffer.

MEDICAL ASPECTS--DIAGNOSIS (GENERAL)

- 2160 INGRAM, T. T. S. The new approach to early diagnosis of handicaps in childhood. *Developmental Medicine and Child Neurology*, 11(3):279-290, 1969.

The early identification of handicapped children is proposed to offer the best chance of ameliorating their condition and requires the recognition of "high risk" infants whose subsequent management offers new challenges. The suspicion that the child will be "at risk" of abnormalities can arise before conception, during pregnancy, labor, and delivery, or in early childhood. Before conception, the presence of a family history of genetic disease, the finding of cytological or biochemical variations in the parents, or the presence of suspicious characteristics in the parents themselves allow informed concern for the offspring. During pregnancy, labor, and delivery, many conditions or events (such as prematurity, rubella infections, and polyhydramnios) are danger signals. After birth, many clinical and laboratory signs may increase the suspicion of the physician long before clinical syndromes are apparent. It is important to follow such cases closely so that diagnosis can be made at the earliest moment. Even when the diagnosis is still obscure, it will often be possible to treat observed deficits in development with good results. This procedure of frequent observation of the infant will, of course, arouse parental anxiety; this has to be recognized and the parents supported throughout the process. (87 refs.)
W. J. Klein.

Department of Child Life and Health
University of Edinburgh
17 Hatton Place
Edinburgh 9, Scotland

- 2161 CHORUS, A. M. J. Fysiognomiek in het bijzonder t.a.v. zwakzinnigen (Concerning physiognomy and its relationship to mental retardation). *Tijdschrift voor Zwakzinnigheid en Zwakzinnigenzorg*, 5(2):33-46, 1968.

Some symptoms of MR can be observed in the face and expression of the MR, such as a total lack of expression in the SMR and a partial lack in the TMR, discrepancies in facial expression and body movement, and inability of

the eyes to focus on a particular object. Although the science of physiognomy has fallen into disrepute, it is helpful in the initial diagnosis of MR. Factors in this diagnosis are general body structure, structure of the head, and most specifically facial expressions. (10 refs.) - S. L. Hamerley.

Rijksuniversiteit Leiden
Leiden, Holland

- 2162 LITTLEFIELD, JOHN W. Prenatal diagnosis and therapeutic abortion. *New England Journal of Medicine*, 280(13):722-723, 1969. (Editorial)

Prenatal diagnoses of several inherited disorders are now possible with amniotic fluid examination and culture of desquamated fetal cells. Although procedures for chromosomal analysis, enzyme assay, and metachromatic staining are still in development, they will someday offer a definitive genetic diagnosis. The possibility of therapeutic abortion should then be available to the mother who does not wish to bear a defective child. (6 refs.)
E. L. Rowan.

No address

- 2163 PAINE, RICHMOND S. Early recognition of neuromotor disability in infants of low birthweight. *Developmental Medicine and Child Neurology*, 11(4):455-459, 1969.

Children of low birthweight are specially liable to cerebral palsy, mental retardation, and other disabilities. In trying to diagnose or foresee such disabilities in an infant, the motor signs and reflex findings should be assessed repeatedly and compared with those of normal infants of similar age, judged by the estimated time of conception. (22 refs.) - *Journal summary*.

Department of Neurology and Neurosurgery
Children's Hospital of the District of Columbia
Washington, D. C.

- 2164 LIND, T., & HYTTEN, F. E. Relation between birth-weight and rupture-delivery interval. *Lancet*, 1(7601):917-918, 1969.

In a series of 243 multigravida with Rh isoimmunization, the interval between surgical rupture of the membranes and delivery was found to be related to the birthweight of the infant rather than to its stage of gestation. The proportion of infants delivering within 24 hours of rupture of the membranes rose progressively with weight from nil for babies under 4 pounds to over 90% for those of 8 pounds or more. The length of gestation, within weight groups, had no effect. The majority who were to deliver did so within 12 hours. (3 refs.) - *Journal summary*.

Princess Mary Maternity Hospital
Newcastle Upon Tyne NE2 3BD
England

- 2165 LUBCHENCO, LULA O. Assessment of gestational age and development at birth. *Pediatric Clinics of North America*, 125-145, 1970.

Methods for estimating gestational age of infants are: calculations based on mother's last menstrual period; obstetric clinical estimates based on events during pregnancy, physical examination of the mother, and the growth of the fetus; clinical assessment based on physical and neurologic characteristics of the infants; and laboratory estimates based on clinical and chemical tests of the infant. Each method has its own drawbacks. The fourth method would be the most precise one; however, the techniques remain to be developed. The interrelations between gestational age, birth-weight, mortality, morbidity, intrauterine growth retardation, and postnatal development need further elucidation. (16 refs.) - *L. S. Ho*.

University of Colorado Medical Center
Denver, Colorado 80220

- 2166 MUNDEL, G., & LEVY, I. Intrauterine growth retardation. *Harefuah*, 75(3): 108-111, 1968.

In assessing several methods of determining gestational age in infants admitted to a prematurity ward, it was found that head circumference, posture, and elbow recoil correlated well with gestational age. In addition, skin structure and color, ear shape and consistency, breast and nipple development, and

plantar creases appear to assess gestational age more reliably than signs used by other investigators. Combinations of such signs may lead to a more accurate gestational age assessment. (41 refs.) - *A. Huffer*.

Assaf Harofe Government Hospital
University of Tel Aviv Medical School
Tel Aviv, Israel

- 2167 NELIGAN, GERALD, & PRUDHAM, DEREK. Norms for four standard developmental milestones by sex, social class and place in family. *Developmental Medicine and Child Neurology*, 11(4):413-422, 1969.

Norms for the developmental milestones of sitting and walking unsupported, using single words and using sentences, as defined in writing, have been worked out from data concerning the whole child population of the City of Newcastle upon Tyne born during the year 1961. The distributions for all 4 milestones are markedly skewed, to approximately the same extent, and the results are presented in terms of percentiles in a way which is potentially of practical use. The correlations between the ages at which children pass the 2 performance milestones and the 2 verbal milestones are much higher than the correlations across these divisions. The 2 milestones, one motor and one verbal, on which we considered the mothers' evidence most likely to be accurate, were walking and using sentences. These were therefore investigated further. Girls and first-born children show significant advancement in using sentences; children in Social Classes II, IV and V show significant advancement in walking. Lapse of time in terms of years before the mother's history is given markedly reduces the reliability of her evidence. (10 refs.)

Journal summary.

Princess Mary Maternity Hospital
Great North Road
Newcastle Upon Tyne, England

- 2168 NELIGAN, GERALD, & PRUDHAM, DEREK. Potential value of four early developmental milestones in screening children for increased risk of later retardation. *Developmental Medicine and Child Neurology*, 11(4): 423-431, 1969.

Children later excluded from normal school on account of mental defect, cerebral palsy, or deafness, show a significant delay in passing the milestones of sitting and walking unsupported, and using sentences; the use of

single words provides less reliable information. Children later admitted to normal schools show a significant relationship between delay in walking and using sentences and a low IQ at the age of 5 years, which reaches a practical level of specificity when the information for both milestones, based on near-contemporary data, is combined. (7 refs.) - *Journal summary*.

Princess Mary Maternity Hospital
Great North Road
Newcastle Upon Tyne, England

- 2169 SAMAN, NAGUIB A., BRADBURY, JAMES T., & GOPLERUD, CLIFFORD P. Serial hormonal studies in normal and abnormal pregnancy. *American Journal of Obstetrics and Gynecology*, 104(6):781-794, 1969.

Serum human placental lactogen (HPL), human chorionic gonadotropin (HCG), and human growth hormone (HGH) levels for the most part could not be correlated with the fetal status when these hormonal levels in normal and abnormal pregnancies (Rh-immunization, diabetes, and toxemia) were compared. Urinary estriol and pregnanediol in diabetic patients were somewhat higher than in controls; however, these levels had no prognostic value in immunization. In 3 patients, the level of HPL was related to fetal outcome. One was an Rh-sensitized patient in whom HPL levels dropped progressively; the newborn died. The second patient had low-normal HPL levels and delivered an infant with symptoms of placental dysfunction. The last patient also had low and decreasing HPL levels and delivered a small-for-dates infant. More research in this area is indicated. (23 refs.) L. S. Ho.

Department of Obstetrics and Gynecology
University of Iowa Hospital
Iowa City, Iowa 52240

- 2170 ACEVEDO, H. F., STRICKLER, H. S., GILMORE, JAMES, VELA, B. A., CAMPBELL, E. A., & ARRAS, BETTY J. Urinary steroid profile as an index of fetal well-being. *American Journal of Obstetrics and Gynecology*, 102(6): 867-879, 1968.

One hundred and seven normal and abnormal pregnancies were studied for urinary estrogen excretions of the fetoplacental unit (FPU) in the human in order to establish a urinary

steroid profile as an index for normal and abnormal status of the FPU. The urine specimens were determined for total estrogens (TE) by automated fluorometry and for pregnane derivatives by gas liquid chromatography (GLC). The patient was considered normal if the entire clinical, prenatal, labor, delivery, and postpartum course was without complication. Patients were considered abnormal on the basis of either their past clinical history or on the outcome of this study. The GLC technique permitted simultaneous determination of pregnanolone (PL), pregnanediol (PD) and pregnanetriol (PT). Six hundred and seventy-six analyses of each steroid or a total of 2,704 determinations were made in the 14-month study. Urinary PL and PD determinations represent indices of the gestational activity of the placenta or of the corpus luteum of pregnancy in the early stages of gestation. Determinations of TE and PT give indices of the status of the fetal part of the FPU. Low values in all values of TE, PL, PD, and PT forecast poor fetal progress. Such a fetal survey is valuable if an examination is made in each trimester of pregnancy or if serial determinations are made in those cases considered clinical risks. In cases of threatened abortion, no miscarriages occurred in those cases with normal PL and PD urinary levels while those who aborted had abnormally low PL and PD levels. (29 refs.) - F. J. McNulty.

Division of Obstetrics and Gynecology
Allegheny General Hospital
Pittsburgh, Pennsylvania

- 2171 SMITH, VERNON H. The ophthalmological examination of the handicapped child. In: Gardner, Peter, MacKeith, Ronald, & Smith, Vernon, eds. *Aspects of Developmental and Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p. 101-102.

It is difficult to examine a handicapped child because physical or mental handicaps often obscure visual problems, visual capacity fluctuates with general health, and the child may not be relaxed during the examination. Since existing hospital systems are potentially frightening to the child, he may be uncooperative at the time of the appointment. Also, he may have psychological difficulties associated with a visit to a doctor. Recommendations for ophthalmological exams include using a nursery for the exam, concealing the equipment, designing visual objects to resemble toys, and utilizing other experts for consultation. (No refs.) - V. G. Votano.

2172 ZIVIN, L., & MARSAN, C. AJMONE. Incidence and prognostic significance of "epileptiform" activity in the EEG of non-epileptic subjects. *Brain*, 91(Part IV):751-778, 1968.

Epileptiform discharges (EDs) in non-epileptic Ss are useful in diagnosing and placing patients of a given age into disease categories which take into account the developmental rate of the disease and associated CNS complications. The study re-evaluated the incidence and type of epileptiform activity in patients with no history of seizures on the basis of personal case material. Specific factors which play a role in the occurrence of epileptiform activity in such cases were investigated and the literature was reviewed for analogous data and information. In addition, the possible prognostic implications of such activity were assessed. Of 6,497 unselected non-epileptics receiving EEGs, 142 had EDs, and these Ss received clinical and EEG follow-ups for a few months to more than 10 years. There was a high rate of EDs in Ss with congenital and perinatally acquired brain damage, brain neoplasms, cranial operations, MR, biochemical disorders, and treatments with anti-neoplastic agents and/or steroids. Twenty Ss subsequently developed seizures. No relation was found between type and distribution of EDs and seizure susceptibility. No Ss with a 6/second spike-and-wave complex developed seizures; 22.46% of Ss classified as having progressive neurological involvement had seizures with tendencies toward only one attack; and 9% of Ss with static neurological diseases developed recurrent seizures. Prognostic meaning of typical EEG changes, while not being fully established, suggests that greater attention should be paid to the dynamic clinical situation (slowly or rapidly changing neurological state). (111 refs.)
R. K. Butler.

National Institute of Neurological
Diseases and Blindness
National Institutes of Health
Bethesda, Maryland 20014

2173 GLOOR, P., KALABAY, O., & GIARD, N. The electroencephalogram in diffuse encephalopathies: Electroencephalographic correlates of grey and white matter lesions. *Brain*, 91(Part IV):779-802, 1968.

EEG changes found in 32 Ss with diffuse encephalopathies were related to the distribution of the pathological process in cortical and subcortical grey matter, cortical white matter, and cortical and subcortical white

matter. Bilaterally synchronous paroxysmal discharges in the EEG were associated with diffuse cortical and subcortical grey matter lesions, while white matter lesions were associated with continuous non-paroxysmal polymorphous Δ activity. Diffuse encephalopathies involving both grey and white matter showed prominent bilaterally synchronous paroxysmal discharges and continuous non-paroxysmal polymorphous Δ activity. Periodic bilaterally synchronous paroxysmal discharges were found in subacute encephalitis of the Dawson and van Bogaert types and in spongiform encephalopathies of the Jakob-Creutzfeldt or Heidenhain types. The EEG pattern has thus been shown to be useful in the differential diagnosis of diffuse encephalopathies. (49 refs.)
M. G. Conant.

McGill University
Montreal, Quebec, Canada

2174 WOLPERT, SAMUEL M. Dural sinus configuration: Measure of congenital disease. *Radiology*, 92(7):1511-1516, 1969.

Venous and sinus configurations were investigated by angiography in cases of Dandy-Walker syndrome, infratentorial cysts, supratentorial cysts, aqueduct stenosis, and communicating hydrocephalus. The values of the tentorial angle between the slope of the straight sinus and a line joining the nasion with the tuberculum sellae and the ratios of the nasion-inion distance to the inion-posterior lip of foramen magnum distance were determined and compared with those of normal controls. Analysis of the results with respect to the embryology of sinus development suggested that the variations in sinus configuration reflect abnormalities occurring at a stage in fetal development prior to the fixity of the dural venous sinuses to the skull, which occurs in the third fetal month. An exception is communicating hydrocephalus which constitutes a lesion in which the abnormality must have occurred later than the first trimester of fetal life. (17 refs.) - M. G. Conant.

Tufts-New England Medical Center Hospitals
Boston, Massachusetts 02111

2175 STOOL, SYLVAN E., GRAHAM, WILLIAM, & RANDALL, PETER. Velopharyngeal incompetence: Pseudo mental retardation as a consequence. *Clinical Pediatrics*, 8(1):42-46, 1969.

An 8-year-old girl with severe hearing and speech disorders was mistakenly diagnosed as MR; however, careful examination demonstrated

a failure of the soft palate to close the nasopharynx (velopharyngeal incompetence) with subsequent unintelligible, hypernasal speech and chronic otitis media with a secondary conductive hearing loss. Failure of velopharyngeal closure during phonation was demonstrated on lateral neck X-rays. At surgery, a pharyngeal flap was constructed, middle ears were drained, and artificial eustachian tubes were inserted. The dramatic postoperative improvement in speech and hearing pointed up the value of special evaluative studies and avoided the tragic mislabeling of this child as retarded. (12 refs.) - E. L. Rowan.

Children's Hospital of Philadelphia
1740 Bainbridge Street
Philadelphia, Pennsylvania 19146

- 2176 OPITZ, JOHN M., HERRMANN, JURGEN, & DIEKER, HANS. The study of malformation syndromes in man. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 1-10.

A vast number of human malformation (dysmorphogenetic) syndromes exist, but remain clinically and etiologically undefined because of their large number, etiologic heterogeneity, complexity, and variability. Delineation of a specific syndrome progresses from the stage of physical examination to formal genesis to causal genesis. Physical examination or simple observation of anomalies requires the prior establishment of strict anthropometric criteria of normality and population surveys to establish a normal range of variability. A formal genesis syndrome is established by finding similar sets of anomalies in several patients. Variable penetrance and expressivity of the anomaly and heterogeneous causality make this difficult. A causal genesis syndrome (concurrence of causally related anomalies) is established firmly only when the genetic basis has been determined. Multifactorial inheritance and inadequate methods of chromosome examination therefore limit the number of validly established, causally determined syndromes. (22 refs.) - E. L. Rowan.

- 2177 HOEFNAGEL, DICK. Malformation syndromes with mental deficiency. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 11-14.

Malformation syndromes hidden in the literature or observed singly by clinicians might become better known and commonly recognized if physicians interested in such disorders would communicate with each other. The deLange and Prader-Willi syndromes had been described in the foreign language literature while American investigators were still puzzling over clinical examples. Combinations of anomalies such as MR with tibial and fibular bowing and MR, vertical talus, and low ridge count have been documented in individuals and have probably been observed by other investigators. A device such as a prepublication "traveling register" based on case histories might be circulated among investigators so that other cases with similar anomalies might be collected. (12 refs.)

E. L. Rowan.

- 2178 SMITH, DAVID W. Recognizable patterns of malformation in childhood. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 255-270.

Although evaluation, prognosis, management, and genetic counseling are relatively straightforward in malformations based on a single primary defect, the situation is more complex when there appear to be multiple primary major or minor defects in morphogenesis. A "core pattern of defects" is the basis on which a table of 130 disorders was constructed. Groups include ectodermal dysplasias, connective tissue disorders, mucopolysaccharidoses, altered skeletal morphogenesis, chromosomal abnormalities, miscellaneous disorders, hamartoses, and abiotrophies. All but 2 of the syndromes are genetic in etiology, but the physician must exercise care in making a correct diagnosis and should take a careful family history before offering a prognosis or genetic counsel. (53 refs.)

E. L. Rowan.

- 2179 MINTZER, R. A., & REISMAN, L. E. Prenatal chromosome studies: Diagnosis in genetic defects. *Southern Medical Journal*, 62(10):1220-1222, 1969.

Samples of amniotic fluid were obtained by abdominal amniocentesis from pregnant patients with Rh sensitization (40 cases), diabetes (2 cases), possible anencephalic fetus (1 case), and from 10 normal pregnant patients. The fluid was washed with Hanks solution and incubated in tissue culture medium which was changed every 3 days. Fibroblast growth began after 15-21 days and 5-7 days later, when an active monolayer had been established, colchicine was added to arrest the cells at metaphase; they were then fixed, stained, and analyzed. Of the 53 samples, 30 successful cultures were obtained, all of which revealed normal karyotypes. The likelihood of successful culture decreases as the length of gestation increases. The ability to evaluate easily and reliably genetic high-risk patients is clearly available. (7 refs.) - M. G. Conant.

Department of Pediatrics
University of Louisville
School of Medicine
Louisville, Kentucky 40202

- 2180 GARDNER, LYTT I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, 1072 p. \$34.00.

A treatise for students, geneticists, and clinicians on pathology, diagnosis, and therapy of endocrine and genetic diseases in childhood is presented. A special effort is made to provide advice on differential diagnosis and management. The major parts of the book are: normal and abnormal growth patterns, abnormalities of various endocrine glands, and inborn errors of metabolism and chromosomal abnormalities. Many genetic disorders are associated with MR; the most common condition is probably mongolism which occurs in 1/600 births. The overall risk of parents having a second offspring with Down's syndrome is 1-2%. Mongolism can be readily diagnosed on the basis of clinical features alone and medical care for mongoloids is the same as for normal persons. Management of these patients should emphasize socialization, training, and recreation. The less frequently occurring chromosomal abnormalities associated with MR include: the *cri-du-chat* syndrome; 18 trisomy and D_1 trisomy syndromes; deletions of autosomes; Turner's syndrome; Klinefelter's syndrome; and other sex chromosome aberrations. Dermatoglyphics, in some cases, can be helpful in diagnosing chromosomal abnormalities.

Endocrine gland disorders associated with MR include cretinism, congenital goitrous hypothyroidism, and hypoparathyroidism. Metabolic disorders associated with MR are galactosemia, hypoglycemia, phenylketonuria, and homocystinuria. (4,799 refs.) - L. S. Ho.

CONTENTS: Normal and Abnormal Growth Patterns; Pituitary Disorders; Thyroid Disorders; Parathyroid Disorders; Disorders of the Adrenal Cortex; Ontogenesis of the Gonads; Disorders of Sexual Development; Dermatoglyphics in Medical Genetics; Disease States Exhibiting Visible Chromosomal Abnormalities; Disorders of Potassium and Magnesium Metabolism; Hereditary Metabolic Bone Diseases; Disorders of Catecholamine Metabolism; Disorders of Carbohydrate Metabolism; Genetic Defects of Amino Acid Metabolism; Genetic Defects in Protein Metabolism; Disturbances of Serum Lipoproteins; The Lipidoses; Obesity in Children and Adolescents; Cystic Fibrosis; Disorders of Heme and Porphyrin Metabolism; Psychologic Aspects of Endocrine and Genetic Disease in Children; Genetic Counseling.

- 2181 UCHIDA, IRENE A., & SOLTAN, HUBERT C. Dermatoglyphics in medical genetics. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 8, p. 579-592.

Persons with autosomal and sex chromosomal abnormalities often present characteristic dermatoglyphic patterns of digits, palms, and/or soles. Patients with 21-trisomy have a high frequency of simian creases (58.5% compared to 0.9% for controls) and of a single flexion crease of the fifth digit (17.9%) whereas it is extremely rare in the normal population. No significant deviations from normal are found in 21-mosaics. In 18-trisomy, the most striking finding is a single arch pattern on all, or nearly all, digits. About 50% of 18 trisomy Ss have simian creases. Almost all D-trisomic Ss have distal axial triradii and simian creases on the palms, and some have a peculiar arch fibular or a modified arch-fibular-S pattern on the sole. In the *cri-du-chat* syndrome, 44 of 88 cases have either a simian crease or 2 parallel transverse creases and a mean axial triradial distance of 29%. In sex chromosome abnormalities, deviations from normal are not as striking as those in autosomal trisomies. The most significant finding in Turner's XO is the large pattern with high ridge counts, an average of 166.1 compared to 130.4 for normal females. Patients with XXY chromosome complement have

comparatively low digital ridge counts of about 118; normal males have about 145. Patients with XYY have a characteristic ulnar triradius in association with loop carpal, loop radial or arch radial patterns. Statistics of other autosomal and sex chromosomal abnormalities are not clear. External teratogens may also alter the normal dermatoglyphic pattern. (40 refs.) - L. S. Ho.

- 2182 MENSER, MARGARET A., & PURVIS-SMITH, S. G. Dermatoglyphic defects in children with leukemia. *Lancet*, 1(7605):1076-1078, 1969.

The dermatoglyphics of 25 children with leukemia have been compared with those of 100 controls of similar age and racial background. In the leukemia group, there was a suggestion of an increase in the incidence of abnormal palmar creases: Sydney lines were present in 5/25 children with leukemia and in only 7/100 controls; simian lines were seen in 4 patients and in only 6 controls. The fingertip patterns suggested an increased incidence of arch patterns and decreased incidence of ulnar-loop patterns in children with leukemia. Dermatoglyphic defects in these children may mark a genetically determined predisposition to leukemia, or a prenatal teratogen may be responsible for both the dermatoglyphic defects and the leukemogenesis. (24 refs.) *Journal summary*.

Children's Medical Research Foundation
Royal Alexandra Hospital for Children
Camperdown, Sydney, New South Wales
Australia 2050

- 2183 STOUGH, THOMAS R., & *SEELY, J. RODMAN. Dermatoglyphics in medicine. *Clinical Pediatrics*, 8(1):32-41, 1969.

There is a tremendous range of dermatoglyphic patterns within the normal population, but the increased or decreased frequency of normal patterns, the presence of normal patterns

in unusual places, and unusual combinations of normal patterns can support a clinical diagnosis, particularly with regard to chromosomal abnormalities. Ridge patterns and flexion creases develop between the tenth and eighteenth weeks of fetal life and are particularly manifest on the fingertips (as whorls, loops, and arches), the palms (as ridges, axial triradii, and flexion creases) and the soles (particularly the hallucal area). Particular dermatoglyphic patterns have been described in trisomies 21, 18, and 13-15, the *cri-du-chat* syndrome, Turner's syndrome, Klinefelter's syndrome, and the congenital rubella syndrome. The simplicity and inexpensiveness of such analysis and the relative constancy of findings in each disorder establish dermatoglyphics as a valuable ancillary diagnostic tool. (32 refs.)

E. L. Rowan.

*Children's Memorial Hospital
University of Oklahoma Medical Center
Oklahoma City, Oklahoma 73104

- 2184 ALTER, MILTON. Dermatoglyphics in birth defects. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations*. (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 103-122.

Dermatoglyphic patterns are one of many anatomical traits that enable the clinician to diagnose a specific birth defect. Although the ridge patterns on fingers, palms, and soles are most abnormal in chromosomal defects, there are some single gene disorders in which the dermal ridge patterns differ significantly from population norms. Trisomies G, 18, and D₁ show characteristic patterns as do Turner's, Klinefelter's, and the *cri-du-chat* syndromes. In the de Lange, Rubenstein-Taybi, and prenatal rubella syndromes, the clinician may find frequency patterns which differ from the population means. The tabulation of dermatoglyphic abnormalities in each syndrome and frequency distributions of dermatoglyphic patterns in both the normal population and patients with birth defects may enable the physician to make use of this diagnostic tool in clinical practice. (100 refs.) - E. L. Rowan.

2185 WARBURTON, DOROTHY. Comments on dermatoglyphics. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, p. 123-124.

A discrimination can be made between cases of a deleted chromosome 4 and a deleted chromosome 5 on the basis of dermatoglyphics. In chromosome 4 deletion, the total digital ridge count (mean of 70) is abnormally low when compared to chromosome 5 deletion (mean 122) and the normal population (mean 140). A double loop on the thumb in association with arches on the second and third fingers is also common in chromosome 4 deletion, rare in normals, and has not been found in the chromosome 5 deletion cases. (No refs.)

E. L. Rowan.

2186 ZAVALA, CARLOS. Fingertip patterns. *Lancet*, 1(7595):628, 1969. (Letter)

A study of fingertip arches in 500 normal Mexicans (250 of each sex) revealed that 5 had 6 or more arches (1 had 7 arches, and 2 females had 10 arches). This provides additional evidence that 6 or more fingertip arches is not diagnostic of trisomy E. (8 refs.) - A. Huffer.

Genetics Department
Instituto Nacional de la Nutricion
Mexico 7, D. F. Mexico

2187 PRECHTL, H. F. R., THEORELL, K., GRAMSBERGEN, A., & LIND, J. A statistical analysis of cry patterns in normal and abnormal newborn infants. *Developmental Medicine and Child Neurology*, 11(2):142-152, 1969.

Statistical analysis of cry patterns of normal and neurologically abnormal newborn infants during the first 9 days of life indicated that the cry duration and interval were

characteristic of normal Ss; whereas, in those with consistent and transient neurological abnormalities, the cry duration was longer and the interval larger than that of normal infants. Similar results were obtained with crying elicited by discomfort or by pinching. In normal babies, the cry duration and interval became progressively shorter and less variable during the neonatal period. (8 refs.) - L. S. Ho.

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University of Groningen
Groningen
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2188 GRAUSZ, JOHN P. The fetus and the newborn. *Medical Clinics of North America*, 53(5):1051-1062, 1969.

Technical advances in diagnosis and treatment in the fields of fetology, perinatology, and neonatology are reviewed. Although studies on the human fetus are few, many animal models (dogs, cats, rabbits, guinea pigs, mice, and rats) have been used. The functions of the placenta, including transmission of certain substances and circulation, have been extensively investigated. The amniotic fluid can be obtained relatively easily and can be analyzed to diagnose Rh isoimmunization, sex, gross chromosomal aberrations, and fetal maturity. Ultrasound and radiology are 2 techniques which can be used to assess fetal size, position, and anomalies. These techniques have wide applicability in high-risk pregnancies. The fetus is in great danger in the period from the beginning of labor to the start of independent breathing, and 2 methods used to monitor the condition of the fetus are fetal heart rate measurement and determination of acidosis. The idiopathic respiratory distress syndrome in neonates can be arrested or reversed by treatment of hypoxia and acidosis. Neonatal sepsis can be controlled with modern drugs, and hyperbilirubinemia has also been treated in new ways with excellent results. (123 refs.) - M. G. Conant.

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MEDICAL ASPECTS--PREVENTION AND ETIOLOGY (GENERAL)

2189 GOLD, EDWIN M., & STONE, MARTIN L. Total maternal and infant care: A realistic appraisal. *Journal of Public Health*, 58(7): 1219-1229, 1968.

A new total maternal and infant care program for 1,130 socioeconomically depressed women included experts in obstetrics and gynecology, pediatrics, social work, nutrition, and public health nursing. Both in-patient and out-patient care were utilized. Continuity of physician and public health nurse was maintained for both the mother and child. Eighty-seven percent of the women were admitted to the program before the seventh month of pregnancy, and 93% stayed with it to the end of their care. Only 10% were white; the remainder were divided equally between black and Puerto Rican. High-risk factors averaged 1.6/patient, and compared to data from a different health district, the prematurity and perinatal mortality rate were higher. Improvement of these statistics should demonstrate the value of this program. At least one postpartum visit was made by 74% of the women, but only 43% accepted family planning services. Pediatric care was established during the first 5 days of life and 6 to 8 well-baby clinic visits were performed during the first year of life. Infants with birth weights below 2,501 gm constituted 19.9% of the infant population; congenital malformations occurred in 5%, and deaths before 7 days occurred in 45/1,000 live-births. Of 40 live births below 2,001 gm, 37.5% succumbed, and neonatal deaths under 27 days were 48.5/1,000 live-births. Patient education regarding pregnancy, birth, infant care, nutrition, and family planning was continuous from the first visit. Furthermore, medical students and resident physicians gained education by being involved in the program. Finally, while improved health care has been attained, further innovations in the program should improve it, and this type of care could become a reality in all communities. (5 refs.) - E. Gaer.

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2190 RANTAKALLIO, PAULA. Groups at risk in low birth weight infants and perinatal mortality. *Acta Paediatrica Scandinavica*, (Supplement 193), 1969, 71 p.

Variations in birth-weight, gestational length, and intrauterine growth curves were calculated for 11,905 single births (96% of all births) in North Finland during 1966. The mortality rates were lowest for the weight range of 3,500-3,999 gm and the fortieth week of gestation. On the intrauterine growth curves, the mortality rate was lowest from the fiftieth to the ninetieth percentiles, twice that rate from the tenth to the fiftieth percentiles, and 10 times that rate below the tenth percentile. The correlations of maternal biological characteristics and socioeconomic circumstances with low birth-weight rates and perinatal mortality rates were calculated by discriminant function analysis using 47 variables on 1,000 control cases and the following groups: birth-weight less than 2,500 gm, survivals and deaths; birth-weight less than 2,500 gm, deaths; and birth-weights greater than 2,500 gm, deaths. In all analyses, the maternal biological characteristics were the most important factors, especially previous low birth-weight infants, parity, and advancing age. The high risk groups, as determined by probability classification, contained 41% of all infants with birth-weights under 2,500 gm: with increasing probability of risk, the mean birth-weight and mean gestational length decrease. (174 refs.) - M. G. Conant.

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2191 KHAZIN, AIDA F., *HON, EDWARD H., & QUILLIGAN, EDWARD J. Biochemical studies of the fetus. III. Fetal base and Apgar scores. *Obstetrics and Gynecology*, 34(4):592-609, 1969.

Fetal base determinations were made on samples obtained from the presenting part of the

fetus and were studied for their value as indicators of neonatal condition. Correlation coefficients were calculated for fetal base deficits and Apgar scores at 1 and 5 minutes. Of 602 fetal base determinations done on 140 patients, 20% and 6.9% were associated with Apgar scores of 1-6 at 1 and 5 minutes (fetal base deficit <12.5 mEq/L) and 52.7% and 89% were associated with Apgar scores of 7-10 at 1 and 5 minutes, respectively (fetal base deficit >12.5 mEq/L). Of 265 fetal base determinations obtained less than 30 minutes before delivery, 11.2% and 3.8% were associated with Apgar scores of 1-6 at 1 and 5 minutes, respectively (fetal base deficit <12.5 mEq/L) and 65% and 87% were associated with Apgar scores of 7-10 at 1 and 5 minutes (fetal base deficit >12.5 mEq/L). The generally poor reliability of the fetal base deficit values may be attributed to the relatively long time between sampling and delivery and the fetal heart rate present at the time of sampling. (15 refs.) - M. G. Conant.

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2192 GROSS, H., JELLINGER, K., KALTZENBACK, E., & RETT, A. Infantile cerebral disorders: Clinical-neuropathological correlations to elucidate the aetiological factors. *Journal of the Neurological Sciences*, 7(3): 551-564, 1968.

Clinical-neuropathological analyses of 891 infant necropsy cases considered to have mental and neurological defects were made in an effort to elucidate the etiology of infantile cerebral disorders. Cases were included in which infantile cerebral lesions became "clinically evident during the first 3 years." Disorders without specific neuropathological abnormalities (mongolism) were excluded. Clinical findings alone, without medical history, would result in a correct diagnosis in 15% of the cases. With laboratory tests, such as PEG, EEG, and biochemical analysis, this percentage could be increased to 24.2% of this series. Cases of cerebral malformation were most accurately diagnosed. There was a lack of adequate case history data in 1/5 of the cases sampled. Medical and family histories appeared helpful in about 16% of the patients but did not prove beneficial in 60% of the cases. Medical case history data is of more value if it is exact and complete. Under optimal conditions, the clinician will be unable to recognize the problem in more than 1/2 of the disorders. Neuropathological analysis resulted in a correct classification

in about 80% of the cases. The highest success (90% correct diagnosis) occurred as a result of combined clinical and morphological diagnosis with good medical records. Perinatal distress was the cause as well as the result of brain damage in a considerable proportion of cases. (50 refs.) - B. Bradley.

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2193 THE NATIONAL FOUNDATION-MARCH OF DIMES. *The Clinical Delineation of Birth Defects. Part I. Special Lectures.* (Birth Defects Original Article Series, 5[1]), New York, New York, 1969, 32 p. \$3.00.

The first of 5 parts of the Proceedings of the First Conference on the Clinical Delineation of Birth Defects included 3 special papers presented as background for the conference topic of nosology (classification) of disease. Genetic disorders are usually described in terms of phenotype; however, advances are made with the discovery of genotype. Gene regulation (first paper) and gene expression (second paper) are essential to understanding the total phenotype. Linking phenotype with genotype is important not only from the research viewpoint but is also clinically applicable in areas such as genetic counseling and patient management. (99 refs.) E. L. Rowan.

CONTENTS: Recent Advances in Developmental Biology (Ursprung); Developmental Mechanisms Found in Allophenic Mice with Sex Chromosomal and Pigmentary Mosaicism (Mintz); On Lumpers and Splitters, or the Nosology of Genetic Disease (McKusick).

2194 CARTER, C. O. An ABC of medical genetics: VI. Polygenic inheritance and common diseases. *Lancet*, 1(7608):1252-1256, 1969.

Many morphological characteristics or common diseases are not transmitted by single dominant, recessive, or X-linked genes, but are predisposed by genetic variation at several gene loci and no individual mutant genes make a preponderant contribution to the variation. When the gene frequency of several individual alleles at gene loci is greater than 1%, polymorphism is the result. When the characteristic is indirectly the product of gene activity, as in fingerprint ridge patterns and systolic blood pressure, the variation is likely to be continuous and normally

distributed; however, there are polygenic inheritances with a discontinuous character, such as cleft-lip. This situation can best be explained by the fact that polygenic predisposition is normally distributed and that there is a threshold beyond which embryos are at risk of developing the malformation. Beyond the threshold, the development of malformation depends on additional environment-triggering factors. (12 refs.) - L. S. Ho.

No address

2195 URSPRUNG, HEINRICH. Recent advances in developmental biology. In: National Foundation-March of Dimes. *Clinical Delineation of Birth Defects. Part I. Special Lectures.* (Birth Defects Original Articles Series, Volume 5, Number 1.) New York, New York, 1969, p. 5-10.

The current "reductionist" view holds that complex developmental processes can be explained by the interaction of a large number of fundamentally simple events with the genes as the basic unit of activity. The information content of cells is regulated by genes which may vary in amount within cell lines in the same individual. This variation may be explained in terms of selective gene amplification, unequal gene transcription, or differential protein synthesis. Mechanisms such as hormonal action or nucleocytoplasmic interaction may control differential expression of genetic activity. The stability of the differentiated state may be maintained by a built-in regulatory program but may also become lost through contact with other cells. Detailed information awaits the discovery of a model system wherein a gene locus can be observed transcribing its information. (22 refs.) - E. L. Rowan.

2196 McKUSICK, VICTOR A. On lumpers and splitters, or the nosology of genetic disease. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part I. Special Lectures.* (Birth Defects Original Article Series, Volume 5, Number 1.) New York, New York, 1969, p. 23-32.

In classifying diseases (nosology), geneticists must be both lumpers in search of pleiotrophism (multiple phenotypes based upon a single gene effect) and splitters in search of genetic heterogeneity (more than one distinct gene entity producing the same phenotype). Classification represents a search for

genetic entities and an eventual identification of the responsible gene(s). At present, much work involves the separation of entities previously considered to be a single phenotype. Clinical examination suffers in the face of variable genetic expression but is valuable in identifying traits in extensive kindred groups. Genetic methods include modes of inheritance, allelism tests, and linkage studies. Biochemical studies, when applicable, are the most definitive means of establishing genetic heterogeneity. Thus far, approximately 700 gene loci (and possible sites of genetic error) have been established in man; however, the actual number is probably much larger. (54 refs.) - E. L. Rowan.

2197 THE NATIONAL FOUNDATION-MARCH OF DIMES. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, 284 p. \$15.00.

The second part of the Proceedings of the First Conference on the Clinical Delineation of Birth Defects includes 33 papers and 18 case reports on malformation syndromes. Included are descriptions of new syndromes, further case reports, literature summaries, and both "lumping and splitting" of syndromes in the light of more recent investigations. Most contain photographs and tabulations of malformations. Case reports include photographs and pertinent clinical, biochemical, X-ray, and dermatoglyphic findings. The appendix consists of tables covering 130 malformation syndromes with reference to the core pattern of anomalies, presence or absence of MR and growth deficiency, and etiology. This should be invaluable to the clinician interested in the evaluation, prognosis, management, and genetic counsel of parents and Ss with birth defects. (487 refs.) E. L. Rowan.

CONTENTS: The Study of Malformation Syndromes in Man (Opitz, Herrmann, & Dieker); Malformation Syndromes with Mental Deficiency (Hoefnagel); The HHHO or Prader-Willi Syndrome (Zellweger); The de Lange Syndrome (Smith, Berg, & McCreary); Dermatoglyphic Studies in a Rubinstein-Taybi Patient, Her Unaffected Dizygous Twin Sister and Other Relatives (Herrmann & Opitz); The Broad Thumbs Syndrome --Progress Report 1968 (Rubinstein); A Familial Syndrome of Mental Deficiency and Broad Thumbs (Robinow); The RSH Syndrome (Opitz, Zellweger, Shannon, & Ptacek); The Lissencephaly Syndrome (Dieker, Edwards, Zuerlein, Chou, Hartman, & Opitz); Some Facial Syndromes (Gorlin); Hypertelorism, Microtia and

Facial Clefting: A New Inherited Syndrome (Bixler, Christian, & Gorlin); Familial Telecanthus with Associated Congenital Anomalies (Christian, Bixler, Blythe, & Merritt); The BBB Syndrome--Familial Telecanthus with Associated Congenital Anomalies (Opitz, Summitt, & Smith); The G Syndrome of Multiple Congenital Anomalies (Opitz, Frias, Gutenberger, & Pellett); Current Status of a Family Previously Reported with the Oral-Facial-Digital Syndrome (Thuline); Visceral Anomalies in an Infant with the Goldenhar Syndrome (Opitz & Faith); Familial Goldenhar Syndrome (Summitt); A Dominantly Inherited First Arch Syndrome (Herrmann & Opitz); Genetic Considerations in a Sibship of Cyclopia and Clefts (Cohen & Gorlin); Familial Anomalies in the Pierre Robin Syndrome (Opitz); Not all Dwarfed Mandibles Are Alike (Pruzansky); Malformation Syndromes with Eye or Ear Involvement (Fraser); The Lenz Microphthalmia Syndrome (Herrmann & Opitz); The Zellweger Syndrome (Cerebro-Hepato-Renal Syndrome) (Opitz, Zurein, Vitale, Shahidi, Howe, Chou, Shanklin, Sybers, Dood, & Gerritsen); A New Case of the Zellweger Syndrome (Taylor, Zellweger, & Hanson); The C Syndrome of Multiple Congenital Anomalies (Opitz, McCreadie, Smith, & Johnson); The Meckel Syndrome (Dysencephalia Splanchnocystica, the Gruber Syndrome) (Opitz & Howe); The Smith-Lemli-Opitz Syndrome of Retardation, Urogenital and Skeletal Anomalies (Dallaire & Fraser); The Silver Syndrome in Twins (Rimoin); Macroglossia, Omphalocele, Adrenal Cytomegaly, Gigantism and Hyperplastic Visceromegaly (Beckwith); Comment on the Macroglossia-Omphalocele Syndrome (Cohen); The Aniridia-Wilms' Tumor Syndrome (Fraumeni, Jr.); Animal Models in the Clinical Study of Birth Defects in Man (Barrow); Case Reports.

2198 THE NATIONAL FOUNDATION-MARCH OF DIMES.

The Clinical Delineation of Birth Defects. Part III. Limb Malformations. (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, 234 p. \$15.00.

The third part of the Proceedings of the First Conference on the Clinical Delineation of Birth Defects includes 20 articles and 12 case reports on limb malformations. Although some of the articles deal with reports of specific syndromes, most are quite general and provide excellent background information for the clinician new to the field. Discussions of bone defects, limb deficiencies, limb anomalies, associated anomalies, dermatoglyphics, and hand malformations are comprehensive and readable. The case reports include photographs and laboratory data and present concise yet comprehensive descriptions of several dysmorphogenetic syndromes.

This volume is an excellent reference book for clinicians seeking to identify and classify limb anomalies. (516 refs.) - E. L. Rowan.

CONTENTS: Bone Defects of the Limbs--An Overview (Lenz); Congenital Skeletal Limb Deficiencies--A General View (Freire-Maia); Nomenclature and Classification of Limb Anomalies (O'Rahilly); Associated Deformities of the Head and Hands (Pfeiffer); Recessive Acrocephalosyndactyly with Normal Intelligence (Summitt); An Unusual Form of Acrocephalosyndactyly (Herrmann & Opitz); Oto-Palato-Digital Syndrome (Aase); Discussion on Oto-Palato-Digital Syndrome (Gorlin); The F-Form of Acro-Pectoro-Vertebral Dysplasia: The F-Syndrome (Grosse, Herrmann, & Opitz); Psychologic Test Findings in the F-Form of Acro-Pectoro-Vertebral Dysplasia: The F-Syndrome (Trites & Matthews); Associated Acral and Renal Malformations (Dieker & Opitz); Brachydactyly Type C of Bell (Cloherty); A Familial Dysmorphogenetic Syndrome of Limb Deformities, Characteristic Facial Appearance and Associated Anomalies: The "Pseudothalidomide" or SC-Syndrome (Herrmann, Feingold, Tuffli, & Opitz); A Triad of Unilateral Limb and Skin Deformities and Congenital Heart Disease (Falek); The Kuskokwim Syndrome: An Inherited Form of Arthrogyposis in the Alaskan Eskimo (Wright & Aase); Inability to Open the Mouth Fully: An Autosomal Dominant Phenotype with Facultative Camptodactyly and Short Stature --Preliminary Note (Hecht & Beals); Autosomal Dominant Inheritance of Shortening of the Flexor Profundus Muscle-Tendon Unit with Limitation of Jaw Excursion (Wilson, Gaines, Brooks, Carter, & Nance); Dermatoglyphics in Birth Defects (Alter); Comments on Dermatoglyphics (Warburton); Synopsis of Hand Malformations with Particular Emphasis on Genetic Factors (Temtamy & McKusick); Case Reports.

2199 PFIEFFER, RUDOLF A. Associated deformities of the head and hands. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 19-34.

The heterogeneous concepts of head-hand deformities (dyscrasio-dysphalangias) must be replaced by morphologic classes which may then be subdivided by etiologic groups on the basis of strict definition and mode of inheritance. Four classes of head-hand deformities have thus been determined. Chromosomal disorders include the phenotypes of trisomies 21, 18, and 13. Monogenic inheritance is suggested in the Apert, oral-facial-digital, oculo-dental-digital, facial cleft with limb

deformities, split hand/split foot, and Gruber syndromes. Conditions of exogenous origin such as thalidomide embryopathy comprise the third class. The causes of association such as the de Lange, oligodactyly, Rubinstein

Taybi, Pierre Robin, and Franceschetti syndromes are unknown. Despite a seemingly clear etiology, exact timetable of organogenesis does not explain how these anomalies are related. (109 refs.) - E. L. Rowan.

MEDICAL ASPECTS--ETIOLOGIC GROUPINGS

Infections, intoxication, and hemolytic disorders

2200 ROWAN, DIGHTON F., McCRAW, MILDRED F., & EDWARD, R. DONALD. Virus infections during pregnancy. *Obstetrics and Gynecology*, 32(3):356-364, 1968.

Laboratory techniques for diagnosis of suspected virus illnesses during pregnancy are described in the general areas of direct, indirect, and presumptive diagnosis. Direct and indirect diagnoses are most reliable if proper specimens are obtained during acute or convalescent phases of illness. A list is presented of viruses which are associated with cutaneous eruptions. Rubella is the only known viral infection proven as teratogenic; however, fetal infections seem to occur more often in exanthematous viral diseases. Seven case reports illustrate effective use of laboratory techniques. Evidence of probable *in utero* infection (rubella and ECHO 9 viruses) was found in 2 cases. Indirect serological data of a recent Coxsackie B2 and a Coxsackie B4 and adenovirus infection was found in 2 other patients with acute postpartum illness. A need is indicated for multiple serological tests for antibodies to viruses endemic at the time as well as those previously associated with the illness. (22 refs.) - B. Bradley.

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2201 EMBIL, J. A., KRAUSE, V. W., HALDANE, E. V., EASTERBROOK, K. B., & CROSBY, J. M. Congenital cytomegalovirus infection: Clinical, pathological and virological studies of two fatal cases. *Canadian Medical Association Journal*, 101(12):32-40, 1969.

Two infants severely ill at birth as a result of congenital cytomegalovirus infection were born in Halifax, Nova Scotia, in April and July 1968. Both died within 7 weeks. Clinical,

pathological, and virological studies including serology and electron microscopy were made both before and after death. Cytomegalovirus was isolated in tissue culture from urine specimens from both infants during life and from organ tissues, blood and ascitic fluid after death. Large inclusion-bearing cells mixed with the erythrocytes and leukocytes were seen within the lumina of pulmonary blood vessels. A brief general discussion is presented of the various clinical manifestations of congenital cytomegalovirus infection and of the different serum antibody patterns found in these 2 pairs of mothers and infants. (31 refs.) - *Journal summary.*

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2202 FONDU, P., & DE MEUTER, F. Facial palsy, a manifestation of acquired toxoplasmosis. *Helvetica Paediatrica Acta*, 24(2): 208-211, 1969.

A 3-year-old child presented a facial palsy accompanied by angina, adenitis and by the discovery of pleocytosis, EEG alterations and abnormal lymphocytes. The diagnosis of acquired toxoplasmosis was based on serological findings and on elimination of all other detectable causes. The course was favorable without treatment. (7 refs.) - *Journal summary.*

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- 2203 FELDMAN, HARRY A. Toxoplasma and toxoplasmosis. *Hospital Practice*, 4(3):64-72, 1969.

Toxoplasma is a ubiquitous intracellular parasite about whose biology, transmission, and clinical expression very little is known. Although serological surveys show a variable but substantial portion of any population to have evidence of *Toxoplasma* infection, there is very little evidence of clinical disease. Transmission has been shown to occur *via* ingestion of cyst-infected meat and transplacentally. The congenital form of disease is devastating. The diagnostic tetrad includes chorioretinitis, hydrocephaly or microcephaly, psychomotor retardation, and cerebral calcifications; the mortality rate is about 25%. Acquired toxoplasmosis is usually innocuous but is associated with posterior granulomatous uveitis and disseminated disease in individuals with immunological deficits. Diagnostic techniques include a skin test (limited value), indirect hemagglutination test, dye test, complement-fixation test, and indirect fluorescent antibody procedure. The only known treatment (sulfadiazine and pyrimethamine) is largely unsuccessful, and agents which attack encysted *Toxoplasma* are unknown. (No refs.) - E. L. Rowan.

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- 2204 ACETO, THOMAS, JR., BECKHORN, GORDON D., MATTIMORE, JOSEPH M., & KENNY, FREDERIC M. Cortisol secretion in children with varicella or rubeola. *American Journal of Diseases of Children*, 117(3):294-298, 1969.

Children (CA 3 to 10 yrs) with varicella (18 Ss) or rubeola (16 Ss) excreted normal (mean 2.2; range 1.0 to 4.0 mg/sq m body surface/24 hrs) amounts of 17-hydroxycorticosteroids, the principal metabolites of cortisol. Sick children receiving glucocorticoids who are exposed to or develop varicella or rubeola should be given physiologic amounts (12 to 36 mg/sq m/24 hr orally) of cortisone acetate until the skin lesions are healed. (19 refs.)
L. S. Ho.

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- 2205 JEAN-JOSEPH, PAUL, IMPERATO, PASCAL J., SOW, SULEYMAN, HENDERSON, RALPH H., CASEY, HELEN L., & NOBLE, JOHN, JR. A comparison of Edmonston-B and Schwarz measles vaccine in Malian children. *Lancet*, 1(7596):665-667, 1969.

A double-blind controlled trial of the clinical and immunological response of 206 Malian children to Schwarz and Edmonston-B vaccines was conducted. Clinical reactions were greater among those receiving Edmonston-B vaccine. Antibody tests indicate that both measles vaccines are effective immunizing agents when administered at the same time as smallpox vaccination. (14 refs.) - *Journal summary*.

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- 2206 SZIRTES, G., CSOKA, E., & LIPCSAK, M. Chromosome alterations in leucocytes from subacute sclerosing panencephalitis patients. *Nature*, 222(5194):692-693, 1969. (Letter)

The persistence of measles virus in human cells is suggested as the etiology of subacute sclerosing panencephalitis (SSPE). Six patients with SSPE were noted to have high titers of measles hemagglutination-inhibiting antibodies and examination of chromosomal preparations from these patients showed patterns of breakage and pulverization similar to those known to be virus-induced. (9 refs.)
E. L. Rowan.

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- 2207 HORTA-BARBOSA, LUIZ, FUCCILLO, DAVID A., & SEVER, JOHN L. Subacute sclerosing panencephalitis: Isolation of measles virus from a brain biopsy. *Nature*, 221(5184):974, 1969. (Letter)

Measles virus was isolated from the brain of a patient with subacute sclerosing panencephalitis (SSPE) by culturing brain fibroblasts with HeLa cells. HeLa cells are from a continuous cell line known to be susceptible to measles. The primary brain cultures of a child with SSPE were trypsinized and suspended in Earle's minimum essential medium.

Mixed cultures were prepared by combining brain cell suspension with HeLa cell suspension in a 1:2 ratio. After incubation for 5 days, the mixed cultures with HeLa cells showed cytopathogenic effects--syncytial and giant cell formation. Fluids from these cultures were frozen, thawed, and inoculated into HeLa cell monolayers and the same cytopathogenic effects appeared on the fourth to fifth day of incubation. Hemadsorption with 0.6% rhesus monkey erythrocytes was positive in these cultures. (7 refs.) - F. J. McNulty.

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2208 ILLAVIA, S. J., & WEBB, H. E. Maintenance of encephalitogenic viruses by non-neuronal cerebral cells. *British Medical Journal*, 1(5636):94-95, 1969.

Two group B tick-borne arboviruses (Kyasanur Forest disease and Langat viruses) which are highly encephalitogenic to mice were inoculated into tissue culture of mouse and fetal human brain glial elements and survived for a long time without apparent damage to the cultured cells. Langat virus was released from the mouse brain tissue culture cells for 58 days and Kyasanur Forest disease virus was liberated for at least 51 days. It appears that these non-neuronal cells can support viral multiplication without sustaining damage. This may explain the persistence of latent viral infections within the brain and the finding that hypertrophy and proliferation of the glial cells are characteristic of chronic central nervous system disease. (12 refs.) - E. L. Rowan.

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2209 RADL, H. Die Bedeutung der Mumpsmeningitis (The significance of mumps meningitis). *Deutsche Medizinische Wochenschrift*, 94(32):1599-1603, 1969.

Mumps meningitis occurs with a frequency that suggests that it is not a complication, but a primary infection due to neurotropism of the virus. Mumps meningitis seldom leaves lasting disability, in contrast to mumps encephalitis where about 30% of the survivors have sequelae, including paralysis, behavior disorders, blindness, hearing loss or deafness, epilepsy and, rarely, MR. In a study of 342 cases with either mumps meningitis or

encephalitis, it was noted that 14 cases were not diagnosed as mumps because parotid swelling was absent; 20 cases occurred before swelling onset; 31 cases occurred at the time of swelling, and in 277, the CNS disease appeared a few days after swelling. Strict bed rest during acute parotitis appears to prevent development of meningitis. EEGs are useful in determining prognosis, particularly with encephalitis. Lasting cerebral injury in children below age 6 is greater than realized. Of 71 children studied, 11 showed behavior changes and 12 had EEG changes leading to seizure activity. Of the cases observed, 224 were 6 to 10 years old; the youngest, 5 months. The report of a case of mumps in the first trimester of pregnancy with subsequent delivery of a malformed infant with cardiac anomalies and early neonatal death suggests that this area needs attention. (26 refs.) - C. A. Rizvi.

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2210 BRUNELL, PHILIP A., BRICKMAN, ANTHONY, & STEINBERG, SHARON. Evaluation of a live attenuated mumps vaccine (Jeryl Lynn) with observations on the optimal time for testing serologic response. *American Journal of Diseases of Children*, 118(3):435-440, 1969.

Live-attenuated Jeryl Lynn strain mumps vaccine produced a slower rise in serum antibody than had been previously noted. Nevertheless, vaccinees tested at 5 weeks following immunization had higher antibody titers and a higher rate of successful immunization than at 4 weeks, and those with no detectable antibody at 4 weeks had higher antibody titers at 8 months. In 2 separate field trials, the vaccine was given to 2,936 children. The proportion of children who were found to be susceptible prior to immunization decreased with age. By multiplying the proportion of children who were susceptible by the number of vaccinees in each age group, it was estimated that 58.2% of the children were susceptible at the time of immunization. The frequency of temperature elevation in the 2 groups of vaccinees was essentially the same. One group included susceptible children who had a rise in serum antibody following immunization, and the other group contained children who were immune at the time they received the vaccine. (15 refs.) - F. J. McNulty.

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- 2211 Second-trimester rubella also hits hard.
Medical World News, 10(13):44, 1969.

A National Institute of Health study of pregnant women during the 1964 rubella epidemic shows that defective children were born to 17% of the mothers who had had rubella in the first trimester and to 10% of the mothers with rubella in the second trimester. In the latter case, the defects may not be so readily apparent at birth nor so severe, but the child may still be severely handicapped. It is extremely difficult to get an accurate record of such children since there are many cases in which the mother showed no symptoms of having had the disease. (No refs.)

E. F. MacGregor.

- 2212 BEST, JENNIFER M., *BANATVALA, J. E., & WATSON, D. Serum IgM and IgG responses in postnatally acquired rubella. *Lancet*, 2(7611):65-68, 1969.

Sucrose-density-gradient studies on sera from patients recently convalescent from rubella showed that specific IgM could be detected by hemagglutination-inhibition tests for up to 20 days from the onset of illness. This was associated with a significant increase in the concentrations of total IgM but not IgG, the IgM response being maximal 5-14 days after the onset of illness. After a month, only IgG could be detected. Complement-fixation antibodies, even when appearing early, consisted of IgG alone. Serum-total-IgG concentrations showed little variation in healthy patients throughout pregnancy, while IgM concentrations were reduced during the second trimester of pregnancy. The presence of a rubella-infected conceptus did not produce an unusually prolonged IgM response. Reduction in antibody titer in sera after treatment with 2-mercapto-ethanol provided a less reliable method for demonstrating recent infection, since reductions in titer were associated usually with significantly raised total IgM levels. (21 refs.) - *Journal summary*.

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London, S. E. 1, England

- 2213 DOWNIE, JEAN C., & OXFORD, J. S. Persistent rubella virus infection in hamster lung cells. *Journal of General Virology*, 5(1):11-17, 1969.

Cell cultures from the lungs of hamsters which had been infected by the intranasal instillation of Judith or HPV 77 vaccine strains of rubella virus continued to produce active rubella virus for 12 subcultures. In newly

seeded cultures of uninfected cells, there was a lag period of 24 hours in which no growth occurred, while, in infected cells, this lag period extended for 48 hours; however, the final number of cells was the same in both groups and these cells could not be differentiated by light microscopic techniques. The growth of infected cells was not altered by differences in temperature or culture medium, nor was it inhibited by usually virustatic compounds such as amantadine, ammonium acetate, or hydrocortisone sodium succinate. Persistently infected cells were resistant to superinfection with vaccinia and *Herpes simplex* viruses despite an absence of detectable interferon and a rubella infection rate of less than 50% of the cells. The hamster lung cell culture will be a valuable system for more detailed study of the relationship between rubella virus and host cells. (14 refs.) - E. L. Rowan.

University of Sheffield
Sheffield 10, England

- 2214 JUST, M., BURGIN-WOLFF, A., EBNER, R., & RITZEL, G. Rotelnimpfung bei jugenden weiblichen Erwachsenen (Immunization against rubella for young adult women). *Schweizerische Medizinische Wochenschrift*, 98(40):1549-1551, 1968.

A trial vaccination of 232 young adult females with attenuated rubella virus showed the following results: of 175 women with previous immunity, 26% showed a small and 22% a major increase in titer, and all but one S with no previous immunity showed formation of antibodies. Some minor side effects, such as, a mild itching rash and fever, were noted, but, it is felt that these are minor compared to possible birth defects in children whose mothers are exposed to rubella during pregnancy. All young girls should be vaccinated against rubella during high school. (18 refs.) - S. L. Hamerley.

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Romergasse 8, Basel 5, Switzerland CH 4000

- 2215 DETELS, ROGER, KIM, KENNETH S. W., GUTMAN, LAURA, & GRAYSTON, J. THOMAS. Live attenuated rubella virus vaccine given in an orphanage just prior to a rubella epidemic. *Journal of the American Medical Association*, 207(4):709-712, 1969.

Fourteen Chinese preschool children in a Taipei, Taiwan orphanage were immunized with

live-attenuated rubella virus vaccine HPV-77 together with 14 control children; the vaccine behaved as it did with previously tested American children. None of the children given the HPV-77 vaccine had local or systemic reactions, fevers, or clinical rubella, and there was no transmission to the 14 susceptible controls in contact with the immunized children. Simultaneously, while the study was made to confirm the safety of the vaccine in Chinese children, a rubella epidemic occurred in the orphanage. One clinical case of rubella occurred in the 14 children given the HPV-77 vaccine 2.5 months earlier. Unfortunately, a second blood sample could not be obtained from this child, and it is unknown if he had an antibody response. There were 20 cases of rubella among the 55 unvaccinated children between the ages of 1 and 6. The HPV-77 vaccine provided protection against clinical rash. Six cases of clinical rubella occurred in a group of children given HPV-77+3 African green monkey kidney tissue culture vaccine. Five cases had onset within 15 days of the time of vaccine injection and are presumed to have been infected with rubella prior to the time of immunization. The sixth case occurred on the twenty-first day, at the extreme limit of the usual incubation period. (13 refs.)

F. J. McNulty.

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2216 Development of rubella vaccine. *Sight Saving Review*, 38(3):155-156, 1968. (Editorial)

Attenuated rubella virus vaccine development in duck embryo, grivet monkey kidney, or rabbit kidney cell culture of strains Merck (Benoit), Meyer-Parkman (HPV-77), HPV-120, Cendehill, and RA 27/3 are discussed. (No refs.) - F. J. McNulty.

2217 MEYER, HARRY M., JR., PARKMAN, PAUL D., HOBBS, THOMAS E., LARSON, H. ELLIOTT, DAVIS, WILLIAM J., SIMSARIAN, JAMES P., & HOPPS, HOPE E. Attenuated rubella viruses: Laboratory and clinical characteristics. *American Journal of Diseases of Children*, 118(2):155-165, 1969.

The first attenuated rubella virus, high passage virus-77 (HPV-77), produced asymptomatic, noncommunicable immunizing infections in children and adults; however, women in the 20-40 year age group occasionally developed signs of mild rubella with transient arthritis.

Virus could be recovered from throat swabs and, rarely, from the blood of vaccinees who were protected when challenged intranasally with unmodified virus. Vaccinees and persons infected earlier with natural rubella virus sometimes had booster responses after exposure to unmodified virus which indicates that limited subclinical reinfection had occurred. The immunity conferred by the HPV-77 vaccine is relatively long-lasting; antibodies have persisted in vaccinees for 3 years without significant decline in titer. (39 refs.)

M. G. Conant.

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2218 HILLEMANN, MAURICE R., BUYNACK, EUGENE B., WHITMAN, JAMES E., JR., WEIBEL, ROBERT W., & STOKES, JOSEPH, JR. Live attenuated rubella virus vaccines: Experiences with duck embryo cell preparations. *American Journal of Diseases of Children*, 118(2):166-171, 1969.

Vaccination of 265 susceptible children with HPV-77 duck vaccine and 33 susceptible children with Merck-Benoit B level virus provoked serologic responses in 97% and 100%, respectively, with no infection of susceptible contacts. About 40% of susceptible adult women vaccinated with HPV-77 showed mild and transient arthritis-arthralgia. The protective efficacy of B, C, and D level Merck vaccine and of HPV-77 duck vaccine was 100%, and the immunity conferred persisted for one year following HPV-77 and 22 or 32 months following Merck-Benoit B or C level vaccine. The required virus dose for immunization was 40 tissue culture doses of HPV-77. Total compatibility was found for trivalent HPV-77 rubella, Moraten line measles, and Jeryl Lynn mumps virus vaccine and for bivalent rubella-mumps and measles-mumps vaccines. The duck embryo HPV-77 vaccine appears ready for consideration for routine use in human Ss. (14 refs.)

M. G. Conant.

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2219 PRINZIE, ABEL, HUYGELEN, CONSTANT, GOLD, JEROME, FARQUHAR, JOHN, & MCKEE, JAMES. Experimental live attenuated rubella virus vaccine: Clinical evaluation of Cendehill strain. *American Journal of Diseases of Children*, 118(2):172-177, 1969.

The Cendehill strain of live attenuated rubella virus vaccine is highly effective, evoking antibody response in nearly 100% of the

susceptible recipients (subcutaneous doses of 50 TCID₅₀) with no decrease in titer up to 24 months after vaccination. The vaccine produces no significant symptoms, is noncommunicable to susceptible Ss in close contact, and produces no adverse reactions among children and young girls. Minimal signs of arthralgia sometimes occur in adult women. The Cendehill rubella virus vaccine is compatible with the live measles vaccine in a bivalent vaccine. Fetal samples obtained by therapeutic abortion from 10 women vaccinated 18-20 days prior to the scheduled abortion have remained seronegative, suggesting that the attenuated strain does not cross the placental barrier or that it has lost its infectivity for fetal tissues. (20 refs.) - M. G. Conant.

Recherche et Industrie Therapeutiques
Genval, Belgium

2220 PLOTKIN, STANLEY A., FARQUHAR, JOHN D., KATZ, MICHAEL, & BUSER, FRITZ. Attenuation of RA 27/3 rubella virus in WI-38 human diploid cells. *American Journal of Diseases of Children*, 118(2):178-185, 1969.

The RA 27/3 strain of live rubella virus attenuated in WI-38 human diploid cells gives a good antibody response in vaccinees without significant reaction and without spread to contacts. Of 775 seronegative persons receiving the vaccine (500 subcutaneously and 225 intranasally), none have shown serious reactions and virtually 100% of the subcutaneous vaccinations have evoked antibodies. There were no joint symptoms in nearly 150 seronegative women who have received RA 27/3 and nearly 400 controls have remained seronegative despite close contact with vaccinees. (10 refs.) - M. G. Conant.

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2221 McCOLLUM, ROBERT W., RANDOLPH, MARTIN F., BRYNE, EARL B., & HILLEMANN, MAURICE R. Rubella virus vaccine (HPV-77-DE 5): Antigenic and protective efficacy in a community trial. *American Journal of Diseases of Children*, 118(2):186-189, 1969.

A field trial of rubella virus vaccine, HPV-77-DE 5, was carried out on 2,630 children

from 1,230 families during a period of natural rubella activity. In families with 2 or more children, one served as an uninoculated control for the first 4 weeks of the study. This control sibling was then inoculated with the same lot of vaccine as his siblings and observed for an additional 4 weeks. Prevacination serologic studies indicated that 63.3% of the children were susceptible at the time of vaccination compared with an overall conversion rate of 97.6% after vaccination. Clinical rubella disappeared among the vaccinees while it still occurred at a steady rate among the uninoculated controls and the total elementary school population. No immediate or delayed local or systemic reactions were observed, thus establishing the safety of HPV-77-DE 5 in children. (4 refs.)
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2222 GOLDBLUM, NATAN, SWARTZ, TIBERIO A., KLINGBERG, WANDA, GOLDWASSER, ROBERT A., & *KLINGBERG, MARCUS A. Immunization with live attenuated rubella virus vaccine (HPV-77): Clinical and serological results in children and adolescents in Israel. *American Journal of Diseases of Children*, 118(2):190-196, 1969.

A group of 239 female Ss (CA 6-29 yrs) were tested for rubella antibody (75 were seronegative) and were then vaccinated with rubella virus vaccine HPV-77. Seventy initially susceptible Ss and 164 seropositive Ss were followed clinically for 28 days and their sera were tested 42 days after vaccination for rubella hemagglutination-inhibiting antibodies. Of the initially susceptible Ss, 89.9% responded serologically, with the percentage of seroconversions decreasing from 100% in the 6-9 year age group to 75% in the 20-29 year age group. Some susceptible Ss experienced mild clinical reactions (lymphadenitis, fever, and rash) and 5 cases of arthralgia were noted: 4 in a group of 7 women aged 20-29 years and one in a 13-year-old girl, all of whom were vaccinated close to their menstrual period. (17 refs.) - M. G. Conant.

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2223 KARCHMER, ADOLF W., HERRMANN, KENNETH L., FRIEDMAN, JOEL P., SHOPE, THOMAS C., PAGE, E. EUGENE, JR., DRESSLER, MARION S., ARMES, WILLIAM H., JR., & WITTE, JOHN J. Comparative studies of rubella vaccines. *American Journal of Diseases of Children*, 118(2):197-202, 1969.

Clinical and serologic data from 2 field trials of attenuated rubella vaccines on 1,174 Ss in Memphis (Tennessee) and 847 Ss in DeKalb County are presented; the vaccines were HPV-77 passaged 5 times in duck embryo cell culture (HPV-77DE₅), HPV-77 passaged 12 times in dog kidney cell culture (NPV-DK₁₂), HPV-77 passaged 3 additional times in primary green monkey kidney cell culture (HPV-80), and the Cendehill strain of rubella virus. The Ss were divided into 5 groups, 2 of which received a saline placebo, and 3 of which received one of the vaccines by subcutaneous injection. Blood specimens were collected immediately prior to vaccination and 50-55 days later, and all Ss were observed for immediate adverse reactions. Seroconversion rates among the 651 susceptible Ss in the group of 1,174 were 100%, 82%, and 77%, respectively, for HPV-77DK₁₂, HPV-77DE₅, and HPV-80, and for the 400 susceptible Ss in the group of 847, 95%, 89%, and 91%, respectively, for HPV-77DK₁₂, HPV-77DE₅, and Cendehill. There were no immediate adverse reactions after vaccination of susceptible or immune children; however, mild, transient joint symptoms were associated with administration of HPV-77DK₁₂ to susceptible children. (24 refs.) - M. G. Conant.

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2224 SCHIFF, GILBERT M., RAUH, JOSEPH L., & ROTTE, THOMAS. Rubella vaccine evaluation in a public school system. *American Journal of Diseases of Children*, 118(2):203-208, 1969.

A rubella serological surveillance and vaccine evaluation program was conducted in the Princeton School District; the program involved 1,477 elementary school children who received either Cendehill or HPV-77-DK-12 live, attenuated rubella virus vaccine. Controls were children who were susceptible, but absent at the scheduled time of vaccination, or who were immune, or nonparticipating. Pre-vaccination and postvaccination paired blood specimens were collected from 1,258 susceptible children, and the hemagglutination-inhibiting antibody level was determined. Seroconversion occurred in 98.9% of the vaccinated children (both vaccines produced

identical conversion rates), and reactions to both vaccines were minimal. This study indicated that rubella vaccination can be effectively done within the school system. (14 refs.) - M. G. Conant.

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Research
Cincinnati, Ohio 45219

2225 SAIDI, SIMON, & *NAFICY, KIARASH. Subcutaneous and intranasal administration of RA 27/3 rubella vaccine: Alone and in conjunction with live attenuated measles vaccine. *American Journal of Diseases of Children*, 118(2):209-212, 1969.

Forty-five children who were seronegative for measles and rubella were divided into 5 groups and given measles vaccine (Group 1), measles plus rubella subcutaneously (Group 2), measles plus rubella intranasally (Group 3), rubella subcutaneously (Group 4), or rubella intranasally (Group 5). The RA 27/3 rubella vaccine and the Schwarz measles vaccine were used. Seroconversion rates were 6 of 7 in Group 1, 7 of 8 for measles and 8 of 8 for rubella in Group 2, 9 of 10 for measles and 7 of 10 for rubella in Group 3, 12 of 12 in Group 4, and 7 of 8 in Group 5. The low seroconversion rate in Group 3 may be due to the small number of Ss. One of 19 seronegative contacts developed rubella antibody. (13 refs.) - M. G. Conant.

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2226 KATZ, SAMUEL L., LANG, DAVID J., WILFERT, CATHERINE M., FEIGIN, RALPH D., & GOLDFEIN, MICHAEL. Children immunized with HPV-77 rubella vaccine: Additional laboratory observations. *American Journal of Diseases of Children*, 118(2):213-217, 1969.

In a closed population of 13 children (CA 38 mos to 9.5 yrs), 11 received HPV-77 rubella vaccine and 2 were given a sterile tissue culture medium placebo. Laboratory techniques included pre- and postvaccination blood specimens for amino acid determinations, and antibody level; chromosome examination; culture and platelet count; throat swab and urine virus culture. Viruria and viremia were not detected, no significant changes in platelet count were detected, and no patterns of breaks nor constrictions were observed in the chromosomes examined. A reversal of the circadian periodicity of blood amino acids, which

began 24-48 hours after vaccination and persisted for 4-14 days, was observed in all the children vaccinated, but in neither of the controls. (11 refs.) - M. G. Conant.

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2227 COOPER, LOUIS Z., ZIRING, PHILIP R., WEISS, HELENE J., MATTERS, BARBARA A., & KRUGMAN, SAUL. Transient arthritis after rubella vaccination. *American Journal of Diseases of Children*, 118(2):218-225, 1969.

Live, attenuated rubella virus vaccine (HPV-77DK12, HPV-77DE5, or Cendehill 51 strain) was administered to 69 adult men and women and to 317 children. Side effects were insignificant except for joint manifestations, which were indistinguishable from those observed during natural rubella and which occurred in 37 (40%) susceptible adults who received derivatives of the HPV-77 strain and in 32 (25%) of the adults who received the Cendehill strain. The severity of the joint pain ranged from mild to transiently debilitating with tenderness and effusion. Paresthesia was prominent in the adults and was observed in 3% of the 295 children receiving derivatives of the HPV-77 strain; although, the children did not develop arthritis. Joint complaints were generally milder and less frequent with the Cendehill vaccine and were more frequently seen in adult women. (14 refs.) - M. G. Conant.

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2228 WEIBEL, ROBERT E., STOKES, JOSEPH, JR., BUYNACK, EUGENE B., & *HILLEMANN, MAURICE R. Live rubella vaccines in adults and children: HPV-77 and Merck-Benoit strains. *American Journal of Diseases of Children*, 118(2):226-229, 1969.

HPV-77DE5 live attenuated rubella virus vaccine was administered to 35 adult women (CA 22-41 yrs) of whom 34 responded serologically. Eight developed a rash, 5 developed nodes, 9 had malaise and/or anorexia, and 15 developed transient symptoms of arthritis-arthralgia with the proximal interphalangeal joints and the knees most frequently involved. The vaccine caused no clinical reaction when given to susceptible females aged 1-14 years, while 10% of susceptible girls aged 15-17 years

showed mild, clinically unimportant, symptoms of arthritis-arthralgia. Level E Merck-Benoit virus was found to induce inadequate antibody responses in adult women. (21 refs.) M. G. Conant.

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Merck Institute for Therapeutic Research
West Point, Pennsylvania 19486

2229 BOUE, ANDRE, PAPIERNICK-BERKHAUER, EMILE, & LEVY-THIERRY, SOPHIE. Attenuated rubella virus vaccine in women: Clinical trials during the postpartum period. *American Journal of Diseases of Children*, 118(2):230-233, 1969.

Hemagglutination-inhibiting rubella antibody determinations were made in 1,500 women about 8 weeks pregnant, and of the 8% found to be susceptible, 36 were vaccinated with HPV-77 vaccine 1-6 days after delivery. Two months later, 32 of the mothers had seroconverted, while none of the 20 infants tested, including 6 breast-fed babies, showed rubella antibody. One case of transient arthritis was observed. Apparently, postpartum vaccination of mothers is safe for both mother and child. (3 refs.) M. G. Conant.

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Paris 16, France

2230 BRYNE, EARL B., RYAN, JOHN M., RANDOLPH, MARTIN F., & HORSTMANN, DOROTHY M. Live attenuated rubella virus vaccines in young adult women: Trials of Cendehill and HPV-77DE5. *American Journal of Diseases of Children*, 118(2):234-236, 1969.

Twenty-four susceptible young adult women were given either Cendehill strain rubella virus vaccine (12 Ss) or HPV-77DE5 (12 Ss), and blood specimens and throat washings were collected several times during the 35 post-vaccinal days. In each group of 12 women, 11 showed a serologic response with a fourfold or greater increase in hemagglutination-inhibiting antibody, and 6 developed a clinical reaction with transient arthritis reported in 3 women in each group. Virus was isolated from only 3 throat washings taken on postvaccination days 12, 13, and 21 from 3 Ss. No virus was isolated from blood specimens; however, the intermittent pattern of sample

collection may have allowed brief viremias and episodes of pharyngeal shedding to go undetected. (4 refs.) - M. G. Conant.

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2231 DUDGEON, J. ALASTAIR, MARSHALL, WILLIAM C., & PECKHAM, CATHERINE S. Rubella vaccine trials in adults and children: Comparison of three attenuated vaccines. *American Journal of Diseases of Children*, 118(2): 237-242, 1969.

Either HPV-77DE₅, Cendehill strain, or RA 27/3 rubella virus vaccine was administered to susceptible adults in a closed population (Group 1), to susceptible student nurses (Group 2), and to susceptible secondary school children (Group 3). In each group, the Ss were divided at random into 2 subgroups, one of which received a vaccine, while the control subgroup was vaccinated 6 weeks later. Mild symptoms were reported most frequently in Group 2 after vaccination with Cendehill and HPV 77, and most infrequently in Group 3. Joint symptoms, reported most often in adult women, were noted after vaccination with HPV-77DE₅ and RA 27/3. Seroconversion rates were above 95% and one probable case of a contact infection from a Cendehill vaccinee shedding virus was reported. Virus was isolated from nose and throat swabs taken on postvaccination days 2-29 from 7 Ss receiving the Cendehill vaccine, 6 given HPV-77DE₅, and one who received RA 27/3. (6 refs.) - M. G. Conant.

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2232 VAHERI, ANTTI, VESIKARI, TIMO, OKER-BLOM, NILS, SEPPALA, M., VERONELLI, JORGE, ROBBINS, FREDERICK C., & PARKMAN, PAUL D. Transmission of attenuated rubella vaccines to the human fetus: A preliminary report. *American Journal of Diseases of Children*, 118(2):243-246, 1969.

Thirty-five pregnant women scheduled for legal abortion were vaccinated with HPV-77DK₁₂ rubella virus vaccine (32 patients) or with HPV-77DE vaccine (3 patients) and aborted 14-24 days later. Fetal samples, placenta specimens, and maternal specimens (blood, pharyngeal swabs, and uterine cervical swabs) were examined independently in 3 collaborating laboratories. Seroconversion was recorded in all of the 22 initially seronegative women,

2 of whom reported joint manifestations. Rubella virus was recovered from many maternal specimens, from initially seronegative women, from 3 placental specimens, and from 1 fetal kidney. The last isolation was made in 2 of the 3 laboratories; however, attempts at re-isolation from the same tissue and at isolation from other tissues from the same fetus were unsuccessful. This fetus was aborted 24 days after vaccination, the longest interval tested. This study indicates that proper precautions should be observed to avoid inadvertent vaccination of pregnant women. (4 refs.) - M. G. Conant.

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2233 PAVILANIS, VYTAUTAS, CHAGNON, ANDRE, DAVIGNON, LISE, & JONCAS, JEAN. Vaccination of infants with HPV-77 vaccine. *American Journal of Diseases of Children*, 118(2):247-251, 1969.

HPV-77 live rubella virus vaccine was given to 24 institutionalized infants 5-12 months of age (19 were seronegative and 5 had maternal hemagglutination-inhibiting antibodies), and normal tissue culture medium was given to 16 controls (12 of whom were seronegative and 4 had maternal hemagglutination-inhibiting antibodies). Mild symptoms (temperature and rash) occurred in both groups with equal frequency, and none of the controls developed rubella antibodies after 90 days of close contact, despite the fact that 27% of the vaccinees showed pharyngeal shedding of virus. Seroconversion occurred in 16 of 19 susceptible vaccinees. One of the seropositive infants vaccinated showed an increased antibody titer, while the antibody levels of seropositive controls decreased. (11 refs.)
M. G. Conant.

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2234 BERKOVICH, SUMNER, STEINER, PHILLIP, & STEINER, MORRIS. Live rubella virus vaccine in tuberculous children. *American Journal of Diseases of Children*, 118(2):252-257, 1969.

The Cendehill strain of rubella virus vaccine was administered to 18 tuberculous children, 14 of whom were seronegative, while 6 children (4 of whom were seronegative) served as

controls. Seroconversion occurred in all 14 susceptible vaccinees, while antibody levels of the controls remained unchanged. Infection with the attenuated rubella virus did not adversely affect the clinical status of the vaccinees, who all received daily doses of antituberculous drugs, and changes in tuberculin reactivity, unrelated to the vaccine virus, were reported in both susceptible and nonsusceptible vaccinees. The serologic response of the susceptible vaccinees was unaltered by the presence of viral and/or bacterial agents implicated in febrile illnesses which appeared in the ward 24-72 hours after the vaccine was administered. (9 refs.)

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- 2235 BOUE, ANDRE, & CELERS, JOSETTE. Attenuated rubella virus vaccines in children with tuberculosis. *American Journal of Diseases of Children*, 118(2):258-260, 1969.

Twenty-eight tuberculous children (CA 2-5 yrs) were vaccinated with HPV-77 rubella virus vaccine (20 Ss) or with RA 27/3 rubella virus vaccine (8 Ss), regardless of serologic status, while their unvaccinated roommates served as controls. Among the 14 susceptible children given HPV-77 vaccine, 12 seroconverted, while all 5 of the susceptible children given RA 27/3 seroconverted. None of the 11 susceptible controls in the group receiving HPV-77 or the 5 susceptible controls in the group given RA 27/3 seroconverted. There were no clinical symptoms observed which were attributable to vaccination and the clinical status of the tuberculosis disease was unaltered. Incidental cortisone therapy received by 9 of the 16 susceptible contacts did not alter their lack of receptivity to the attenuated rubella virus. (3 refs.) - M. G. Conant.

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Paris 16, France

- 2236 LAMB, GEORGE A. Effect of HPV-80 rubella vaccine on the tuberculin reaction. *American Journal of Diseases of Children*, 118(2):261, 1969.

Seven tuberculin-positive children who were susceptible to rubella were given the HPV-80 rubella virus vaccine and all responded serologically. Four of the children showed decreases in the sizes of their tuberculin reactions, 3 to less than 10 mm in diameter,

while the skin tests of the other 3 remained unchanged. The clinical status of the 6 vaccinated children with active, primary pulmonary tuberculosis was unchanged. (3 refs.)

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- 2237 FURUKAWA, TORU, MIYATA, TAKAO, KONDO, KEIZO, KUNO, KUNIYOSHI, ISOMURA, SHIN, & TAKEKOSHI, TERKO. Clinical trials of RA 27/3 (Wistar) rubella vaccine in Japan. *American Journal of Diseases of Children*, 118(2):262-263, 1969.

A total of 80 seronegative persons, including 37 adult women, was given RA 27/3 rubella virus vaccine. Both children and adults were essentially free of clinical symptoms and all the vaccinees seroconverted. Virus was isolated from 7 of 35 Ss tested, but none of 7 susceptible children in contact with vaccinees developed antibodies. Fifteen women 6-10 weeks pregnant and scheduled for abortion were given RA 27/3 vaccine (10 women) or the Cendehill strain vaccine (5 women), and curettage was performed 7-19 days later. Seroconversion occurred in each of the 7 susceptible women and, in all cases, the products of conception were histologically normal and did not contain rubella virus. (No refs.)

M. G. Conant.

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- 2238 GOLD, JEROME A., PRINZIE, ABEL, & MCKEE, JAMES. Adult women vaccinated with rubella vaccine: A preliminary report on controlled studies with Cendehill strain. *American Journal of Diseases of Children*, 118(2):264-265, 1969.

Cendehill strain rubella virus vaccine was given to 116 adult women (CA 18-50 yrs), 46 of whom were seronegative, while 61 adult women, 30 of whom were seronegative, received a saline placebo. Among the 46 seronegative vaccinated Ss, there were 5 cases of mild to moderate arthritis-arthralgia, compared with 3 cases in the seropositive vaccinated group, 4 cases in the seronegative control group, and 3 cases in the seropositive control group. All of the 31 seronegative vaccinees tested had seroconverted. These studies confirm the low incidence of mild, transient, localized

joint manifestations associated with administration of the Cendehill vaccine. (5 refs.) - M. G. Conant.

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2239 FARQUHAR, JOHN D., & CORRETJER, JORGE E.
Clinical experience with Cendehill rubella vaccine in mature women. *American Journal of Diseases of Children*, 118(2):266-268, 1969.

A total of 290 rubella-susceptible women (CA 17-37 yrs) was given the Cendehill rubella virus vaccine at medical centers in Philadelphia, Pittsburgh, and San Juan (Puerto Rico). An additional 140 seropositive women in San Juan were also vaccinated and constituted a control group. A serologic response occurred in 99% of the Ss with the most prominent side effects being postauricular lymphadenopathy (11%) and mild arthralgia (7%). The control group showed about the same incidence of side effects as the experimental group, suggesting the presence of a placebo effect. (6 refs.) - M. G. Conant.

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2240 SCHIFF, GILBERT M., DONATH, RUDOLF, & ROTTE, THOMAS. Experimental rubella studies: I. Clinical and laboratory features of infection caused by the Brown strain rubella virus. II. Artificial challenge studies of adult rubella vaccinees. *American Journal of Diseases of Children*, 118(2):269-274, 1969.

Administration of the Brown strain of rubella virus to 22 adult males with low or undetectable rubella hemagglutination-inhibiting (HI) antibody produced the typical clinical and laboratory features of rubella in 9 of the 14 Ss with HI antibody levels less than 8, in one of 4 Ss with HI antibody levels of 8, and in none of 4 Ss with HI antibody levels of 16-32. Six of 7 susceptible males vaccinated with HPV-80 live, attenuated rubella virus vaccine developed HI antibody. The vaccinees were challenged 3 months later with the Brown strain virus administered intranasally, and evidence for nasopharyngeal replication of rubella virus was found in 2 vaccinees. Four of 5 Ss who had developed HI antibody following vaccination with Cendehill strain virus vaccine were similarly challenged 3 months later. Although the vaccinees were not contagious, rubella virus was isolated from

pharyngeal swabs taken from 2 of them and one showed a significant rise in HI antibody titer. (8 refs.) - M. G. Conant.

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Cincinnati, Ohio 45219

2241 WILKINS, JEANETTE, *LEEDOM, JOHN M., PORTNOY, BERNARD, & SALVATORE, MARGARET A. Reinfection with rubella virus despite live vaccine induced immunity: Trials of HPV-77 and HPV-80 live rubella virus vaccines and subsequent artificial and natural challenge studies. *American Journal of Diseases of Children*, 118(2):275-294, 1969.

In 4 studies in which HPV-77 rubella virus vaccine was given to 16 children in an isolation unit, and HPV-80 vaccine was given to 18 children, seroconversion occurred in all 16 HPV-77 vaccinees and in 15 of the 18 HPV-80 vaccinees. Viremia was reported in one HPV-77 vaccinee and in 2 HPV-80 vaccinees. There was no spread of HPV-77 vaccine to any of 14 susceptible contacts; however, there was serologic evidence of HPV-80 vaccine spread in 1 of 12 susceptible contacts. Intranasal challenge of vaccinees with the Brown strain of rubella virus (16 Ss) or exposure to natural rubella (8 Ss) resulted in serologic evidence of subclinical reinfection in 12 vaccinees, 11 of whom had received HPV-77 vaccine and one of whom had received HPV-80 vaccine. Four of the 12 reinfected Ss shed pharyngeal virus. Challenge of 8 HPV-80 vaccinees with the Brown strain followed by their exposure to 6 rubella-susceptible children 2 days later produced subclinical reinfection in one of the vaccinees, but in none of the contacts. (20 refs.) - M. G. Conant.

*University of Southern California
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Los Angeles, California 90033

2242 DETELS, ROGER, GRAYSTON, J. THOMAS, KIM, KENNETH S. W., CHEN, KUNG-PEI, GALE, JAMES L., BEASLEY, R. PALMER, & GUTMAN, LAURA. Prevention of clinical and subclinical rubella infection: Efficacy of three HPV-77 derivative vaccines. *American Journal of Diseases of Children*, 118(2):295-300, 1969.

A total of 5,994 boys in grades 1-4 were vaccinated; 1,290 with HPV-80 live rubella virus vaccine, 1,082 with HPV-77DE₅ vaccine, 887 with HPV-77KD₁₂ vaccine, and 2,735 with Salk polio vaccine as a placebo, 1-2 months before

a rubella epidemic reached its peak in Taiwan. The efficacy of the vaccine, calculated as the reduction in attack rate in the vaccine group as compared to the placebo group, was 90-95% for all 3 vaccines with the protective effect beginning on the twelfth day and reaching a maximum by the twenty-first day. Rubella virus was isolated at least once from 55% of 82 Ss who had received placebo and from 42% of the vaccinees between 30 and 50 days post-vaccination, with fewer isolations/vaccinated boy than isolations/placebo boy. Apparently, the vaccine reduced viral shedding and presumably viremia. Antibody titers were higher in placebo boys with infection than in vaccinees. (11 refs.) - M. G. Conant.

Conant.

Publications Office

Naval Medical Research Unit Number 2

Box 14, APO San Francisco, California 96263

2243 BEASLEY, R. PALMER, DETELS, ROGER, KIM, KENNETH S. W., GALE, JAMES L., LIN, TUN-LUNG, & GRAYSTON, J. THOMAS. Prevention of rubella during an epidemic on Taiwan: HPV-77 and RA 27/3 rubella vaccines administered subcutaneously and intranasally HPV-77 vaccine mixed with mumps and/or measles vaccines. *American Journal of Diseases of Children*, 118(2):301-306, 1969.

A double-blind, placebo-controlled efficacy trial of HPV-77 and RA 27/3 strain live, attenuated rubella virus vaccines was conducted on 1,462 susceptible male students in grades 1-4 in southern Taiwan during an epidemic in 1968. Eleven groups were given either HPV-80, HPV-77DE₅, or RA 27/3 vaccine administered subcutaneously or intranasally, HPV-80 in combination with mumps and/or measles vaccines, or mumps or measles vaccine alone (placebo). Attack rates in the vaccine and control groups were the same for the first 17 days post-vaccination. Then, the attack rate for the control group was 15.6%, compared with 1% or less for groups given each type of rubella vaccine subcutaneously, whether alone or in combination with mumps and/or measles vaccines. Intranasal administration of HPV-80 induced no immunity while similar administration of RA 27/3 reduced the attack rate to about 7%. No complications were observed in any vaccinee. (13 refs.) - M. G. Conant.

University of Washington
School of Medicine
Seattle, Washington 98105

2244 Immunization of man against rubella. *American Journal of Diseases of Children*, 118(2):307-321, 1969. (Discussion)

Studies on children in Switzerland, Jamaica, Costa Rica, North Carolina, St. Louis, and Nashville confirmed the efficacy, nontransmissibility, and safety of both the Cendehill and the HPV-77 live rubella virus vaccines. The Cendehill vaccine evoked an immune response with no unpleasant side effects in susceptible adult women in Italy, while vaccination of 11 susceptible infants (CA 6-11 mos) with HPV-77 vaccine evoked a serologic response in 8 of them. Administration of HPV-77 vaccine to pregnant women scheduled for therapeutic abortion has produced no evidence for the presence of rubella virus in placental and fetal tissue, but the question of the effect of rubella vaccines on the human fetus is still unanswered, because the susceptible child is more sensitive than tissue culture systems for the detection of rubella virus. The question of frequency of reinfection after vaccination and the possibility of differences in antigenic make-up among the rubella virus strains are considered. (No refs.) - M. G. Conant.

2245 SCHIFF, GILBERT M. Titered lots of immune globulin (Ig): Efficacy in the prevention of rubella. *American Journal of Diseases of Children*, 118(2):322-327, 1969.

Experimental rubella studies on adult male volunteers were conducted to determine the effect of titered lots of immune globulin (Ig) on the prevention of rubella viremia. In 5 Ss given high-titered (4,096/0.1 ml) Ig 24 hours after intranasal exposure to Gilchrist strain of rubella virus, rubella viremia as well as clinical and laboratory signs of infection were absent, while 5 controls developed the typical signs of rubella. In 6 Ss given low-titered (256/0.1 ml) Ig, serologic evidence for infection was present in 4, and 3 showed viremia. The serum hemagglutination-inhibiting antibody titer of 8 Ss not infected with rubella virus was unchanged by the administration of high-titered Ig. (12 refs.) M. G. Conant.

Christ Hospital Institute
of Medical Research
2139 Auburn Avenue
Cincinnati, Ohio 45219

2246 BECK, EARL S. Review of studies with inactivated rubella virus. *American Journal of Diseases of Children*, 118(2):328-333, 1969.

Virus preparations can be made inactive by either β -propiolactone-ultraviolet light or γ -irradiation. Serologic responses (hemagglutination-inhibiting and serum neutralizing antibodies) were obtained in monkeys inoculated with both inactivated preparations; however, antibody levels obtained with the former tended to decrease over a period of time, and with the latter, one inoculation was insufficient to evoke an immune response. The possibility of preparing sub-unit inactivated vaccines is mentioned. (18 refs.)
M. G. Conant.

National Institute of Allergy
and Infectious Diseases
National Institutes of Health
Bethesda, Maryland 20014

2247 MURRAY, RODERICK. Biologics control of virus vaccines. *American Journal of Diseases of Children*, 118(2):334-337, 1969.

The development of a live virus vaccine depends upon knowledge of the origin of the strain, the specificity of the strain with respect to the causative agent of the disease, degree of attenuation, safety, stability of properties, noncommunicability, long-lasting immunity, and efficacy. The degree of attenuation is related to the average level of antibody response and will, therefore, be dependent on the protective antibody level required. Uniform properties of the rubella virus are obtained after 5 passage cultures. Methods of safety testing include long-term evaluation of vaccinees, administration to school age children instead of to the group directly threatened (women of childbearing age), and the explicit statement of possible side effects (arthralgia and transient arthritis). (No refs.) - M. G. Conant.

Division of Biologics Standards
National Institutes of Health
Bethesda, Maryland 20014

2248 HOPPS, HOPE E., PARKMAN, PAUL D., & MEYER, HARRY M., JR. Laboratory testing in rubella vaccine control. *American Journal of Diseases of Children*, 118(2):338-346, 1969.

Marker tests (*in vitro* and *in vivo*) can be applied to potency testing and virus identification tests for licensed live rubella virus

vaccines. HPV-77 live rubella virus vaccine in rabbit kidney cell cultures produces a unique cytopathic effect after 8 days and induces plaques in the same cell culture. HPV-77 induces interferon at low levels *in vivo* and also *in vitro*, as do Merck-Benoit C level vaccine and the Cendehill strain. HPV-77 vaccination induces antibody formation in rhesus monkeys and is not transmitted to the products of conception, while vaccination with a low passage virus produces widespread distribution of the virulent virus, even in the products of conception. Both the seed virus and tissue culture substrate should be carefully screened for the presence of contaminants, especially when new tissue culture systems are used for virus vaccine production (27 refs.) - M. G. Conant.

*National Institutes of Health
Bethesda, Maryland 20014

2249 BUYNACK, EUGENE B., LARSON, VIVIAN M., McALEER, WILLIAM J., MASCOLI, CARMINE C., & HILLEMAN, MAURICE R. Preparation and testing of duck embryo cell culture rubella vaccine. *American Journal of Diseases of Children*, 118(2):347-354, 1969.

The duck embryo cell culture system is useful for attenuating rubella virus virulence for man and for propagating the virus for vaccine use because the duck is relatively free of viral diseases and of neoplasia. No extraneous agent originating in the duck embryo has been found in any duck embryo cell culture in tests using cell cultures, experimental animals, electron microscopic examinations, and immunofluorescent studies. Tumors produced by oncogenic viruses were undetected in tests for resistance-inducing factor and group-specific, complement-fixation tests for avian leukosis or by inoculation of newborn hamsters and ducks. Finally, inoculation of 18,000 children has produced no untoward effects which confirms previous estimates of the safety and applicability of duck embryo cell-culture rubella vaccine. (26 refs.)
M. G. Conant.

*Merck Institute for Therapeutic Research
West Point, Pennsylvania 19486

2250 MUSSER, SAMUEL J., & HILSABECK, LARRY J. Production of rubella virus vaccine: Live, attenuated in canine renal cell cultures. *American Journal of Diseases of Children*, 118(2):355-361, 1969.

Four strains of rubella virus (Gilchrist 13, Gilchrist 48, HPV-77, and HPV-120) grew easily

and rapidly in canine renal cell cultures. Parallel series of vaccines derived from HPV-77 strain and passaged 11 times in canine renal cell culture showed different immunogenic response in monkeys with one group showing 90-100% seroconversion compared with no detectable seroconversion in another group. The difference was attributed to overmodification occurring between passages 1 and 3. Susceptible monkeys and children were inoculated with a single lot of rubella virus vaccine, and antibody response was determined 8 weeks later. The immune response to rubella vaccination is greater in children than in monkeys. (13 refs.) - M. G. Conant.

Philips Roxane
2621 North Belt Highway
St. Joseph, Missouri 64502

2251 HUYGELEN, CONSTANTINE, SIGEL, MICHAEL M., ZYGRAICH, NATHAN, PEETERMANS, JULIEN H., COLINET, GERARD, LEYTEN, ROGER, RAUPP, WILLIAM G., PINTO, CARL A., GARG, SARYU G., BOYLE, JOHN J., & HAFF, RICHARD F. Safety testing of rubella virus vaccine (Cendehill strain): Preparation in primary rabbit kidney cells. *American Journal of Diseases of Children*, 118(2):362-366, 1969.

Experiments indicate that primary rabbit kidney (PRK) cells are suitable as substrates for live, injectable, attenuated rubella virus vaccine. *In vitro* tests, including microscopic observation of PRK cultures, passages in cell cultures, hemadsorption tests, interference tests, electron microscopy, and immunofluorescence, demonstrated no cytopathic, interfering, hemadsorbing, or infectious agent in PRK cell cultures. Oncogenicity studies, inoculation of thymectomized newborn mice, rats, and hamsters with PRK, and transplantation of rabbit kidney cells to the autologous host, produced no tumors or deaths. The use of kidneys from rabbits kept in a homogeneous colony under pathogen-free conditions and constant surveillance is an additional safety factor. (14 refs.) - M. G. Conant.

Recherche et Industrie Therapeutiques
Rue de l'Institut
Rixensart, Belgium

2252 TINT, HOWARD, & ROSANOFF, EUGENE I. Production and testing of rubella virus vaccine: Prepared on WI-38 cell cultures. *American Journal of Diseases of Children*, 118(2):367-371, 1969.

The WI-38 diploid cell strain from human fetal lung is attractive as a substrate for viral

propagation. The cells can be maintained for a limited number of passages in the diploid mode prior to senescence and can be subcultured in expanding cell numbers. Specific long-term safety tests performed on uninoculated control cells include: tests for cytopathic effect on primary cell cultures; aerobic and anaerobic tests for *Mycoplasma*; tests for hemadsorption; observation for viral, bacterial, and fungal growth; cytology of stained preparations; karyology; and heterotransplantation in hamsters. Rubella virus strains can be propagated in the WI-38 diploid cell strain with ease and consistency. (17 refs.) - M. G. Conant.

Wyeth Laboratories
Box 8299
Philadelphia, Pennsylvania 19101

2253 Gamma globulin prophylaxis; inactivated rubella virus; production and biologics control of live attenuated rubella virus vaccines. *American Journal of Diseases of Children*, 118(2):372-381, 1969. (Discussion)

The administration of γ -globulin to 33 pregnant women exposed to rubella after development of rash had no effect on the incidence of rubella defects in the offspring, while treatment of 1,870 cases within 5 days of exposure reduced the incidence to that expected in the average population. The recognition of adventitious viruses in primary cell cultures requires long incubation times for slow and masked viruses. Better methods of cell observation than cytopathic effect observation and hemadsorption are needed. After administration of HPV-77 vaccine cultivated in dog kidney cells, 5 cases of serious reaction have occurred, but at the present time, there is no definitive evidence that there is any cause and effect relationship between the vaccine and allergic phenomena such as anaphylaxis, angioneurotic edema, or urticaria. Use of WI-38 human diploid cells as a substrate for viral propagation, although desirable for obtaining a standardized cell seed system, is potentially hazardous for it may contain an undetected leukemia virus. (No refs.) - M. G. Conant.

2254 Panel discussion on future of rubella virus vaccines. *American Journal of Diseases of Children*, 118(2):382-396, 1969. (Discussion)

Plans for mass vaccination with rubella virus vaccine and the question of priorities were discussed with some participants favoring

first vaccinating prepubertal children, thereby reducing the virus reservoir and the risk to their mothers. This group would be relatively easy to reach through schools. Others favored starting with vaccination of the high-risk group including women in the child-bearing years, female teachers and nurses, and all children under the age of 20 or so. This plan would be complicated by the possible presence of undetected pregnancies, medical-legal questions which would arise if a woman gave birth to a congenitally defective child after receiving the vaccination, and the difficulty in reaching women in the child-bearing years. This latter problem could be circumvented by vaccination of women in the postpartum period, but the risk of early pregnancy would still be present. The question of vaccinating all women or only the susceptible ones, which would involve widespread use of hemagglutination-inhibiting antibody determination, a fairly difficult test, was also discussed. It was generally felt that the most desirable method of vaccine distribution would be to vaccinate all prepubertal children. (No refs.) - M. G. Conant.

2255 COGHLAN, JOYCE D., & BAIN, A. D. Leptospirosis in human pregnancy followed by death of the foetus. *British Medical Journal*, 1(5638):228-230, 1969.

Maternal infection with leptospirosis (Weill's disease) late in pregnancy was probably responsible for a fetal death, although no organisms were recovered from the abortus. The fetus had died at least 2 days before delivery, however, and tissue autolysis had begun. The mother had a very high agglutination titer to serotype *Leptospira canicola* as did her husband, the farm dogs, and several pigs. A febrile illness during pregnancy in an area where leptospirosis is endemic (such as Scottish pig farms) should make this disease suspect, and antibiotic treatment should be instituted in order to reduce the risk of abortion. (12 refs.) - E. L. Rowan.

Department of Bacteriology
University of Edinburgh
Edinburgh 8, Scotland

2256 OH, SHIN JOONG. Cerebral paragonimiasis. *Journal of the Neurological Sciences*, 8(1):27-48, 1969.

Although the lung is the primary site of human infection with *Paragonimus westermani*, the brain is the most frequent extrapulmonary site. In Korea (*Paragonimus* is endemic in

the Far East and Southeast Asia), 62 cases of cerebral paragonimiasis were found--primarily in young people who had ingested raw crayfish. The typical course of disease appeared to be an acute (but usually mild) meningoencephalitis followed by chronic illness. The most common symptoms were seizures, headache, visual disturbances, unilateral weakness, mental deterioration, and nausea and vomiting. Physical signs included mental deterioration, hemiplegia, homonymous hemianopsia, optic atrophy, and hemihypesthesia. Diagnosis of *Paragonimus* infection is made by a positive skin test, a positive complement-fixation test, or by demonstration of the ova in stool, sputum, gastric washings, or tissue. In the cerebral form of infection, the spinal fluid complement-fixation test and colloid gold curve are abnormal. Chest X-rays were abnormal in 80% of patients (a cysto-nodular lesion was characteristic) and intracranial calcifications (some of the "soap bubble" type) were noted in 50%. Pneumoencephalography generally revealed subcortical atrophy and EEGs showed generalized abnormalities. Bithional is the treatment of choice in acute and subacute cerebral paragonimiasis but is ineffective in chronic inactive cases. Surgical intervention is indicated in "tumorous" forms of the disease. (53 refs.) - E. L. Rowan.

Mayo Box 335
University of Minnesota Medical Center
Minneapolis, Minnesota 55455

2257 DEBROISE, A., DEBROISE-BALLEREAU, C., SATGE, P., & REY, M. La trypanosomiasis africaine du jeune enfant (African trypanosomiasis in a young child). *Archives Françaises de Pédiatrie*, 25(6):703-720, 1968.

African trypanosomiasis was diagnosed in 151 Ss in Dakar, and 14 (9%) of these cases were children under the age of 5 years. Fever is the most frequently observed clinical symptom and prominent neurological signs include disturbances of consciousness and sleep, hypotonia, and psychomotor retardation. Laboratory findings include lymphatic meningitis, significantly increased serum IgM, and the appearance of IgM in the cerebral spinal fluid (CSF). Diagnosis is based upon the identification of the parasite in the blood or CSF. African trypanosomiasis occurs in children as a subacute infection, as an acute disease progressing rapidly to death, or as a disease with deceptive symptomatology. Drugs of choice include the arsenical compounds, mel-W or trimelarsan, alone or in combination with

corticotherapy. Satisfying results have been obtained with all except the acute forms of the disease. (18 refs.) - M. G. Conant.

Clinique des Maladies Infectieuses
Faculte de Medecine
Dakar, Senegal, Africa

2258 SANDERS, DORIS Y., & GARBEE, H. WESLEY.
Failure of response to ampicillin in
Hemophilus influenzae meningitis. *American Journal of Diseases of Children*, 117(3):331-333, 1969.

A 21-month-old boy with *Hemophilus influenzae* meningitis failed to respond after 48 hours of intravenous therapy with the recommended dosage of ampicillin despite an apparent susceptibility demonstrable by both disk method and tube dilution antibiotic susceptibility tests. After treatment with chloramphenicol and a tap of a subdural effusion, the patient recovered without apparent sequelae. Sequstration of organisms at the site of effusion may have prevented effective diffusion of the ampicillin. It must be emphasized that the response of the patient is the final test of drug effectiveness and the physician must continue to evaluate both clinical and bacteriologic responses during, as well as prior to, therapy. (10 refs.) - E. L. Rowan.

Department of Pediatrics
Bowman Gray School of Medicine
Winston-Salem, North Carolina 27103

2259 MANIOS, SOTIRIS G., LIPIRIDOU, OLGA, ZAPHIRIOU, JOANES, DANGA, ERIPHILI, & ZERVANOU, EPHI. Meningococcal meningitis: Comparison of ampicillin and combined sulphadimidine penicillin G treatment. *Scandinavian Journal of Infectious Diseases*, 1(2): 85-89, 1969.

Children with meningococcal meningitis were treated either with ampicillin (76 Ss) or the conventional sulfadimidine-penicillin G regimen (83 Ss). There was no significant difference either in the mortality rate or incidence of sequelae in the 2 groups and recovery as evidenced by the time of defervescence, bacteriologic response, and return to normalcy of cerebrospinal fluid values was the same in both. A benign rash developed in 11 patients on ampicillin and in 3 on combined therapy; however, the latter was also responsible for 3 cases of hematuria, 1 of thrombocytopenia, and 1 of jaundice. Ampicillin represents a real advance in management

because of its efficiency and freedom from serious side reactions. Deaths in this series resulted from Waterhouse-Friderichsen syndrome (5 Ss) and coma (1 Ss) so that the next treatment advance must be in supportive measures rather than in the discovery of a new antibiotic. (16 refs.) - E. L. Rowan.

Hospital for Infectious Diseases
199, Konitsa Street
Thessaloniki, Greece

2260 FOX, HOWARD A., HOGEN, PATRICIA A., TURNER, DOROTHY J., GLASGOW, LOWELL A., & CONNOR, JAMES D. Immunofluorescence in the diagnosis of acute bacterial meningitis: A cooperative evaluation of the technique in a clinical laboratory setting. *Pediatrics*, 43(1):44-49, 1969.

Immunofluorescent antibody examination of the cerebrospinal fluid was compared with Gram staining and culture in the identification of *Neisseria meningitidis*, *Diplococcus pneumoniae*, and *Hemophilus influenzae*--the 3 species responsible for most cases of acute bacterial meningitis in childhood. In a sample of 287 patients, there were no significant differences among the 3 methods of examination in the frequency and reliability of identification. The fluorescent technique was of some advantage when there was partial antimicrobial treatment prior to hospitalization (drug inhibited and non-viable bacteria) and when there were relatively few organisms in the cerebrospinal fluid. This technique was done rapidly and no false positives were observed. Fluorescent antibody study is not applicable in neonatal meningitis which is usually Gram-negative in origin with the large number of bacterial serotypes making such study impractical. Equipment is costly and commercial agents unreliable. The technique may be a valuable adjunct to diagnosis in facilities with pre-existing equipment and trained technicians and where a large volume of cerebrospinal fluid studies would make such examination practical. (9 refs.) - E. L. Rowan.

Mount Sinai School of Medicine
New York, New York 10029

2261 BELSEY, MARK A. CFS glutamic oxaloacetic transaminase in acute bacterial meningitis. *American Journal of Diseases of Children*, 117(3):288-293, 1969.

A correlation was found between elevated cerebral spinal fluid (CSF) glutamic oxaloacetic transaminase (GOT) levels and complications including subdural effusions, CNS

sequelae, persistent altered state of consciousness, seizures, and death in acute bacterial meningitis. The clinical course was uncomplicated in 48 of 57 patients with GOT levels lower than 24 units, whereas complications developed in 30 of 38 patients with GOT levels higher than 24 units. The initial CSF GOT was not a reliable index of prognosis for patients who had developed meningitis several days before hospitalization. GOT value was a more specific measure than the CSF glucose or CSF protein level. The elevated GOT level could not be related to pleocytosis. (14 refs.) - L. S. Ho.

Tulane University
1430 Tulane Avenue
New Orleans, Louisiana 70112

2262 SYMMERS, W. ST. C. Primary amoebic meningoencephalitis in Britain. *British Medical Journal*, 4(5681):449-454, 1969.

Primary amoebic meningoencephalitis is caused by amoebae of the genera *Naegleria* and *Hartmannella* (*Acanthamoeba*), which ordinarily are free-living saprophytes. The infection may be acquired from fresh water--for example, while bathing--the amoebae invading the nasal mucosa and reaching the meninges and brain along the olfactory nerve filaments. The disease is designated "primary" to distinguish it from meningocerebral infection caused by the parasitic amoebae, particularly *Entamoeba histolytica*, which invade the central nervous system only as a result of dissemination in the blood stream from lesions in other parts of the body. During histological reappraisal of old specimens in a medical museum in London an instance of amoebic meningoencephalitis histologically indistinguishable from the published cases has been found. The specimen dates from 1909. The patient was said to be from Essex. What may have been another case, seen in Northern Ireland in 1937, is also described briefly. These observations may indicate that this disease occurs in the British Isles. Primary amoebic meningoencephalitis should be considered in the differential diagnosis of every case of acute meningitis. (28 refs.) - *Journal summary*.

Histopathology Laboratory
Charing Cross Hospital
London W. C. 2, England

2263 NICOLOPOULOS, D. MATSANIOTIS, N., KATAMIS, C., & ATHANASSIADES, T. Post-vaccinal complications: Report of 45 cases. *Helvetica Paediatrica Acta*, 24(4):378-389, 1969.

Forty-five cases are reported of post-vaccinal complications manifested within 20 days of a successful smallpox vaccination in a mass campaign in Greece during January, 1962. The types of complication observed were encephalitis (10 cases), thrombocytopenic purpura (4 cases), allergic purpura (2 cases), afebrile convulsions (4 cases), hyperpyretic convulsions (5 cases), nephritis (3 cases), vaccinia generalista (8 cases), eczema vaccinatum (5 cases), inoculation (3 cases), and hypotonia, stupor, and spasmus rotatorius (one case). Complete recovery followed in all cases except for one death and one case of minor neuromuscular sequelae in a 5 who had encephalitis. Complications representing hypersensitivity or allergic reactions were treated symptomatically, while other complications representing viremic complication were treated etiologically. (11 refs.)

M. G. Conant.

"St. Sophie's" Children's Hospital
Athens 608, Greece

2264 UZNOV, G., BOZHINOV, S., & GEORGIEV, IV. K probleme progressivnyushchikh panencefalitov (The problem of progressive panencephalitis). *Zhurnal Nevropatologii i Psikiatrii*, 69(3):321-328, 1969.

Of 96 cases with progressive panencephalitis followed longitudinally, 13 had a chronic disease course with survival from 5 to 12.5 years. Clinical features of the chronic form of the disease can be differentiated into 2 groups; there is a small number of Ss with decerebrate rigidity and a larger group with significant improvement in muscular tone. Cases of remission have occurred with some restitution of intellectual functions; however, the disease appears to be able to recur at any time. It is hypothesized that Dawson's Pette-Doring's, and van Bogaert's encephalitis are all forms of the same disorder, and the etiological agent is a virus related to the rabies virus. (43 refs.) - M. D. Nutt.

No address

2265 WAGNER, H. P., TONZ, O., & GREYERZ-GLOOR, R. D. Congenital lymphoid leukaemia: Case report with chromosomal studies. *Helvetica Paediatrica Acta*, 23(6):591-610, 1968.

A female infant showed hepatomegaly, pancytopenia with hemorrhagic manifestations, and petechiae on the thighs for the first 2 weeks of life after which generalization into a case of congenital lymphoid leukemia occurred. An apparently complete remission was induced by treatment with corticosteroids and 6-mercaptopurine; however, the disease reappeared and efforts to induce a second remission with prednisone and vincristine were unsuccessful. The girl died at age 4 months, and the autopsy revealed leukemic infiltrations in the kidneys, heart, lungs, spleen, liver, bone marrow, and lymph nodes. Inconclusive chromosome studies suggested hyperdiploid modes and the beginning of a clonal evolution. The reported cases of congenital lymphoid leukemia (7 connatal, 6 neonatal, and 4 in mongoloid children) are summarized. The disease affects males more often than females, hemorrhagic and/or nodular skin lesions and hepato- and/or splenomegaly are the most prominent clinical features, the disease is not uniformly fatal in mongoloid children, and death is usually due to infection and/or hemorrhage. In only one case has a cure been reported (a mongoloid child who is still alive at 4.5 yrs). (43 refs.)

M. G. Conant.

Swiss Center for Clinical Tumor Research
Tiefenau
Bern, Switzerland

2266 COSTILOE, TERESA M. Ordinal position in sibship and mother's Rh status among psychological clinic patients. *American Journal of Mental Deficiency*, 74(1):10-16, 1969.

The possible etiological role of Rh encephalopathy in mild psychological deficit was investigated in a psychological clinic through study of ordinal position in family and mother's Rh status. Since Rh-affected cases show a characteristic ordinal position distribution differing from unselected children's, the distribution for patients with Rh-negative mothers was compared with that of their siblings on the assumption that the distribution should not differ if the patients were unaffected. An overrepresentation in later ordinal positions was found for study patients

but not for patient controls and was interpreted as evidence for Rh encephalopathy as etiology of psychological impairments. (21 refs.) - *Journal abstract*.

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Midwest City, Oklahoma

2267 BORRONE, CARLA, & BRICARELLI, FRANCA DAGNA. Survival of donor's lymphocytes in new-born infants submitted to exchange transfusion. *Helvetica Paediatrica Acta*, 24(2):192-197, 1969.

Lymphocyte survival times of transfused blood were studied in 4 newborn infants with severe icterus: 3 girls and 1 boy, submitted to exchange transfusion with blood given by male donors. The sex chromosomes were used as markers. In the first 3 cases, lymphocytes with 46,XY karyotype were found. In the fourth case, the marker was formed by a different length of the Y chromosome; the boy actually had, like his father, a long Y. It was possible to prove the survival of donor's cells, up to the fifty-second day after transfusion. The importance of these findings is stressed, since transfused adult lymphoid cells may provoke in the host immunological reactions, like final destruction of donor's lymphocytes; implantation of transfused lymphocytes, causing troubles similar to "runt disease"; or improved tolerance to grafts of tissues coming from the same donor. (8 refs.) - *Journal summary*.

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Genoa, Italy

2268 BRYANT, E. C., HART, GERALD D., CAIRNS, DAVID, GAMARRA, JORGE A., DE VEBER, L. L., HOLLAND, C. G., & HUTCHISON, J. LAWRENCE. Clinical evaluation of Rh₀(D) immune globulin (human) in Canada. *Canadian Medical Association Journal*, 101(12):82-83, 1969.

During 1966, clinical trials were conducted in 3 Canadian centers to determine the safety and efficacy of Rh₀(D) immune globulin (human) in preventing isoimmunization by the Rh₀(D) antigen in Rh-negative women delivering ABO-compatible Rh-positive infants. The candidates were randomly divided into control and treated groups; the treated mothers received an intramuscular injection of 300 µg of anti-Rh₀(D) within 72 hours of delivery. Follow-up antibody screening tests were conducted on the sera of all patients 6 to 9 months postpartum. Of the 175 control patients, 11 or

6.2% became actively immunized to the Rh antigen, whereas complete protection against maternal Rh immunization was observed in the 191 treated patients. (4 refs.) - *Journal summary*.

Department of Obstetrics and Gynecology
Toronto East General Hospital
Toronto, Ontario, Canada

2269 BISHOP, G. J., & KRIEGER, VERA I. One millilitre injections of Rh₀(D) immune globulin (human) in prevention of RH immunization: A further report on the clinical trial. *Medical Journal of Australia*, 2(July 26):171-174, 1969.

An intramuscular injection of 1 ml of Rh₀(D) immune globulin (1,200 µg anti-D globulin) within 72 hours after delivery of an ABO compatible, Rh-positive baby prevented the development of anti-D antibodies in 95 Rh-negative mothers for at least 6 months after delivery. Among the 131 control mothers, 11 (8.4%) demonstrated antibodies in follow-up tests. There seemed to be no consistent relationship between parity and/or time after delivery in the development of such antibodies. Sensitization apparently occurs as a result of transplacental hemorrhage and immune globulin injected into the mother successfully destroys these fetal cells. (11 refs.) - E. L. Rowan.

386 Albert Street
East Melbourne, Australia 3002

2270 USATEGUI-GOMEZ, MAGDALENA, & STEARNS, SUSAN. Comparative study of the Rh-D antibody titres of amniotic fluids and corresponding maternal sera in Rh-D sensitized pregnancies. *Nature*, 221(5175):82-83, 1969. (Letter)

The titer of maternal Rh antibodies is not reflected proportionally in the degree of fetal erythroblastosis or the titer of antibodies in the cord blood or amniotic fluid. This lack of correlation probably results from individual differences in the maternal-fetal barrier (placental factors). Amniotic fluid studies in 33 Rh-D sensitized pregnancies demonstrated that the amniotic fluid titer was never higher than the maternal titer and that it represented a more reliable index of the severity of hemolytic disease than did the level of maternal antibody. (38 refs.) E. L. Rowan.

Putnam Memorial Hospital
Bennington, Vermont 05201

2271 ROBERTSON, J. G. Management of patients with Rh isoimmunization based on amniotic fluid examination. *American Journal of Obstetrics and Gynecology*, 103(5):713-722, 1969.

A plan for the management of patients with Rh isoimmunization is proposed based on examination of 616 amniotic fluids from 251 patients over 3 years. The fluids are scanned spectrophotometrically and the optical density values at 375, 450, and 525 mµ are transferred to semilog graph paper and a straight line is drawn between the values at 375 and 525 mµ to obtain the expected value at 450 mµ. The difference between this number and the actual value can be used as a guide for determining the time at which the initial amniocentesis is carried out, the frequency of repeat tests, the time at which delivery is indicated, and the indications for intrauterine transfusion. The proposed plan emphasizes practical management rather than prediction of the outcome. (21 refs.) - M. G. Conant.

Simpson Memorial Maternity Pavilion
and Royal Infirmary
Edinburgh, Scotland

2272 CASSADY, G., & BARNETT, R. Acid-base and gas tension studies of the amniotic fluid in human gestation. *Biologia Neonatorum*, 14(3-4):251-263, 1969.

Acid-base and gas tension studies have been determined in 142 amniotic fluids from 107 pregnancies, most of which were complicated by Rh (D) sensitization, diabetes, toxemia, and fetal distress. Mean values for pH of 7.15 ± 0.09 (s.d.), PCO₂ of 47 ± 5.3 mm Hg (s.d.) and PO₂ of 43 ± 19 mm Hg were observed. These parameters were not affected by maternal age, race, nature, or severity of pregnancy complication or fetal condition. Significant linear changes in amniotic fluid pH and PCO₂ were observed with advancing gestational maturity, confirming previous suggestions of progressive accumulation of PCO₂ and [H⁺] as pregnancy advances. Although these parameters have not been of clinical use in the diagnosis of fetal compromise in the present series, the data suggest their potential usefulness in the estimation of fetal maturity. (34 refs.) *Journal summary*.

Department of Pediatrics
University of Alabama Medical Center
Birmingham, Alabama 35233

2273 LEVI, A. J., GATMAITAN, Z., & ARIAS, I.

M. Deficiency of hepatic organic anion-binding protein as a possible cause of non-haemolytic unconjugated hyperbilirubinaemia in the newborn. *Lancet*, 2(7612):139-140, 1969.

The biochemical defect responsible for neonatal non-hemolytic unconjugated hyperbilirubinemia ("physiological" jaundice) in man is believed to be immaturity of uridine diphosphoglucuronyl transferase. An alternate hypothesis is that neonatal jaundice results from defective hepatic uptake of bilirubin due to deficiency of an intracellular organic anion-binding protein. The development of 2 hepatic cytoplasmic organic anion acceptor proteins, Y and Z, was investigated in guinea pigs of various ages. Relative deficiency of Y, the major binding protein, was seen in fetal and newborn animals. (21 refs.)

Journal summary.

Department of Medicine
Albert Einstein College of Medicine
Bronx, New York 10461

2274 ACKERMAN, BRUCE D. Infantile pyknocytosis in Mexican-American infants.

American Journal of Diseases of Children, 117(4):417-423, 1969.

Seven Mexican-American infants with unexplained hyperbilirubinemia were found to have abnormal erythrocytes (pyknocytes or "burr cells") on peripheral smear. Two of these infants required exchange transfusions, and despite this, one appears to have developmental retardation secondary to kernicterus. In no case was there a positive Coomb's test, ABO blood group incompatibility, glucose-6-phosphate-dehydrogenase deficiency, or toxic exposure. The disease process appeared self-limiting. An extracorporeal factor was suspected because the morphologic abnormality was present within hours of exchange transfusion when most cells were of donor origin. Infantile pyknocytosis occurred with a minimum frequency of 1/250 Mexican-American births and should be suspected in all cases of neonatal hyperbilirubinemia. Because of the rapid rise in bilirubin concentration, the level should be followed closely and more liberal criteria adopted for exchange transfusion. (15 refs.) - E. L. Rawan.

Department of Pediatrics
University of California
Irvine, California 92664

2275 HYMAN, CAROL B., KEASTER, JACQUELINE, HANSON, VIRGIL, HARRIS, IRWIN, SEDGWICK, ROBERT, WURSTEN, HELMUT, & WRIGHT, ANN ROSE. CNS abnormalities after neonatal hemolytic disease or hyperbilirubinemia: A prospective study of 405 patients. *American Journal of Diseases of Children*, 177(4):395-405, 1969.

Follow-up studies on 405 children with hemolytic disorder of the newborn and 9 children with hyperbilirubinemia of other etiology showed that 15% had one or more of the following CNS abnormalities: sensori-neural hearing loss (SNHL); athetosis; strabismus; seizures; minimal cerebral dysfunction (MCD) syndrome; and miscellaneous problems (impaired mentality, psychotic behavior, or spontaneous nystagmus). Only SNHL and athetosis were significantly associated with high bilirubin level (higher than 20 mg/100 ml). MCD and auditory rote memory and visual perception difficulties had a suggestive association with bilirubin levels. The incidence of CNS abnormalities was 12% among infants with only mild or no neurologic abnormalities, 29% among those with moderate abnormalities, and 100% among those with marked abnormalities. (25 refs.) - L. S. Ho.

4650 Sunset Boulevard
Los Angeles, California 90027

2276 KEASTER, JACQUELINE, *HYMAN, CAROL B., & HARRIS, IRWIN. Hearing problems subsequent to neonatal hemolytic disease or hyperbilirubinemia. *American Journal of Diseases of Children*, 177(4):406-410, 1969.

Seventeen of 405 children with hemolytic disease of the newborn or hyperbilirubinemia were found to have sensorineural hearing loss (SNHL). Eleven of them had high bilirubin levels. The audiograms showed that the hearing loss was most marked at high frequencies. The behavioral characteristics of these patients were similar to those with hearing loss of other etiologies. SNHL can occur without other central nervous system disorders. (14 refs.) - L. S. Ho.

*Department of Hematology
Childrens Hospital of Los Angeles
4650 Sunset Boulevard
Los Angeles, California 90027

- 2277 YEUNG, C. Y., & FIELD, C. ELAINE. Phenobarbitone therapy in neonatal hyperbilirubinaemia. *Lancet*, 2(7612):135-139, 1969.

The effect of phenobarbitone therapy on neonatal jaundice was tested in 210 jaundiced newborn Chinese babies. The serum-bilirubin levels after treatment in 93 randomly selected babies were compared with that of 117 controls receiving no drug. In 53 controls, the serum-bilirubin continued to rise to high levels requiring exchange transfusion but only 4 babies in the treated group subsequently required an exchange. Statistically significant reduction of bilirubin levels was achieved by phenobarbitone therapy in babies with ABO incompatibility, glucose-6-phosphate dehydrogenase deficiency, cephalhematoma, and non-specific causes. These findings are of considerable importance to the Chinese baby whose immature liver function results in a high incidence of hyperbilirubinemia. (20 refs.) - *Journal summary*.

Department of Pediatrics
University of Hong Kong
Hong Kong, United Kingdom

- 2278 RAMBOER, CARLOS, THOMPSON, R. P. H., & WILLIAMS, ROGER. Controlled trials of phenobarbitone therapy in neonatal jaundice. *Lancet*, 1(7602):966-968, 1969.

Controlled trials were designed to determine the effect of phenobarbitone on neonatal hyperbilirubinemia. In one series, treatment of mothers alone with 60 mg/day from the thirty-second week of pregnancy was shown to be effective. The mean plasma-bilirubin in the babies of treated mothers was 4.4 mg/100 ml on the third day of life, compared with 8.2 mg/100 ml in babies of control mothers. There was only one plasma-bilirubin above 10 mg/100 ml in the babies from the treated group, compared with 6 from the control mothers. In the other series, treatment of either normal or low-weight babies starting immediately after birth, but beforehand, was less effective. Side-effects were slight in all series. (21 refs.) - *Journal summary*.

King's College Hospital
London, S. E. 5, England

- 2279 KOPITO, LOUIS, BRILEY, ANN M., & *SHWACHMAN, HARRY. Chronic plumbism in children: Diagnosis by hair analysis. *Journal of the American Medical Association*, 209(2):243-248, 1969.

The determination of lead in scalp hair is a valuable diagnostic aid in chronic or mild lead intoxication particularly when the other clinical or laboratory evidence is of questionable diagnostic quality. This continuously growing tissue accumulates and stores lead for long periods and may be used for estimating the time and duration of the exposure. Hair is easy to obtain, store, transport, and analyze and may provide a practical means for finding little children who may have been exposed to lead. (14 refs.) - *Journal summary*.

*300 Longwood Avenue
Boston, Massachusetts 02115

- 2280 FREEMAN, R. Chronic lead poisoning in children: A review of 90 children diagnosed in Sydney, 1948-1967. *Australian Paediatric Journal*, 5(1):27-35, 1969.

Data on 90 children from New South Wales for a period of 20 years indicated that lead poisoning was responsible for 11 deaths as well as severe neurological or renal problems in 30% of the group. Most patients had an elevation of blood lead and/or excretion of lead in urine as well as hematological and radiographic changes. The incidence of cases appeared constant until a major increase of 23 cases was found at the Prince of Wales Hospital in 1965. Seventy percent of the cases ranged from 1 to 3 years. Diagnostic evaluation should include data relating to pica history and analysis of sibling behavior. Ingestion of lead paint from walls and outside areas is the leading cause of lead poisoning. Children often did not show specific symptoms but were lethargic and irritable with some gastrointestinal and neurological difficulties. The most reliable test for this group was analysis of lead in the urine. Early diagnosis and treatment are of major importance since often there are no pathognomonic conditions or physical indications. These data from New South Wales indicate that lead poisoning continues to be a health hazard. (12 refs.) - *B. Bradley*.

Prince of Wales Hospital
Sydney, Australia

- 2281 GRAHMANN, H., & REIMER, F. Psychopharmakotoxische Encephalopathie (Encephalitis caused by toxic doses of psychopharmaceuticals). *International Pharmacopsychiatry*, 1(2):126-128, 1968.

Observation of 3 mentally ill Ss after withdrawal of high dosages of psychopharmaceuticals, such as Proxypandy, Reserpine, and Butyrophenon, showed an increase in motor disturbances and possible semi-permanent or permanent damage to the thalamus. Some symptoms of this damage were vague pains and discomforts, overeating, and in the case of one female, the growth pattern of male pubic hair. The general mental and physical condition of all 3 Ss deteriorated with the reduction of medication. (3 refs.) - S. L. Hamersley.

Universitätsnervenklinik
Niemannsweg 147
Kiel, West Germany D 2300

- 2282 SCHOLZ, B., EGGERS, H., KULZ, J., WAGNER, D., & KYANK, H. The physical and mental development of children of mothers with toxemia of pregnancy. *German Medical Monthly*, 13(10):487-491, 1968.

One-hundred-three children (CA 3-6 yrs) born of mothers suffering from toxemia of pregnancy were studied to determine delayed effects. Height, weight, head circumference, neurological findings, carpal radiographs for skeletal maturity, psychological status, and EEG were studied. Nineteen abnormal Ss were found including 8 MRs. The majority of the abnormalities were found among children of mothers with severe preeclampsia or secondary toxemia of pregnancy with hypertension and proteinuria. Other abnormalities were detected in those Ss with retarded intrauterine

growth. Early delivery in these conditions is emphasized. (27 refs.) - W. Asher.

Universitäts-Kinderklinik
Doberaner Strasse 142
X-25 Rostock 1, East Germany

- 2283 KHERA, K. S., & CLEGG, D. J. Perinatal toxicity of pesticides. *Canadian Medical Association Journal*, 100(4):167-172, 1969.

The effects of pesticides on man and animals over the period from gestation to weaning have been studied primarily in avian eggs and small mammals. Organochlorine pesticides, especially DDT, reduce embryonic viability but are not known to possess teratogenic potential. DDT residues do cross the placenta and are found in maternal milk. Organophosphate, cholinesterase-inhibiting insecticides such as parathion have been found to cause malformations and increase neonatal mortality. Some other fungicides and herbicides are teratogenic to laboratory animals. Avian studies are difficult to generalize to man because birds are non-placental and phylogenetically remote from man. In small mammals, extremely large doses are necessary to induce changes, the route of administration is not related to human intake and these animals have a different genetic, metabolic, and placental constitution. The number of species affected by any one pesticide is limited. It is, therefore, unlikely that any one pesticide now in use will cause damage to the perinatal human at normal exposure levels; however, as new compounds and possibly synergistic combinations are developed, constant vigilance must be maintained and research for an "ideal" laboratory animal continued. (67 refs.)

E. L. Rowan.

Food and Drug Directorate, Research
Laboratories
Department of National Health and Welfare
Ottawa 3, Ontario, Canada

Trauma or physical agents

- 2284 ROBERTSON, ANN MARIE, & *CRICHTON, JOHN U. Neurological sequelae in children with neonatal respiratory distress. *American Journal of Diseases of Children*, 117(3):271-275, 1969.

Thirty-three infants with low birth-weight and respiratory distress syndrome (RDS) were compared to controls for neurological abnormalities; neurological abnormalities and abnormal mental development were more frequent in infants with RDS. These differences were not statistically significant. The infants with RDS were compared to control infants for neurological abnormalities, intelligence, EEG findings, and ophthalmological status at the University of British Columbia. Forty-two of the 66 children were of normal neurological state and intelligence. The remaining 24 children were abnormal: 14 RDS and 10 control children. Six RDS and 4 control children had cerebral palsy; 6 RDS and 3 control children had minimal cerebral dysfunction; 2 RDS and 4 control children had cerebral dysfunction; 2 RDS and 2 control children were MR. In some children the handicaps were multiple. Abnormalities on eye examination were found in 6 children with RDS and in 4 control children. The mean IQ for the RDS group was 89.1 and the control group 88. The EEG findings were normal in 41 patients, 17 RDS and 24 controls; borderline in 10 patients, 7 RDS and 3 control children; and abnormal in 7 patients, 4 RDS and 3 control children. The various abnormalities were consistently more frequent in children with RDS than in those without RDS, but not statistically significant. (10 refs.) - F. J. McNulty.

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- 2285 GOMEZ, P. C. W., NOAKES, M., & BARRIE, HERBERT. A prognostic score for use in the respiratory-distress syndrome. *Lancet*, 1(7599):808-810, 1969.

Fifty cases of the respiratory-distress syndrome were awarded a prognostic score retrospectively. This was based on 15 clinical and biochemical observations made over the 6-hour period after admission. All infants were

admitted within 8 hours of birth. The prognosis could be classified as good (score <20), fair (20-30), or virtually hopeless (31-41). Only minimal supportive therapy was necessary for those who scored less than 15, while all 14 infants with scores over 30 died. (30 refs.) - *Journal summary*.

Fulham Hospital
London W. 6, England

- 2286 SAVIGNONI, P. G., BUCCI, G., CECCAMEA, A., MENDICINI, M., SCALAMANDRE, A., & ORZALESI, M. N. Intravenous infusion of glucose and sodium bicarbonate in hyaline membrane disease: A controlled trial. *Acta Paediatrica Scandinavica*, 58(1):1-9, 1969.

Twenty-four newborns with hyaline membrane disease (HMD) treated with intravenous infusion of glucose and sodium bicarbonate showed an improved survival curve when compared with 24 controls. Ninety-five newborns (birth-weights 1.25 to 2.50 kg) with respiratory distress syndrome (Silverman score higher than 2) and abnormal lung X-ray findings were admitted within the first 24 hours of life. In 60 babies, reticulo-granularity (RG) was shown on the chest film; 48 of these Ss were included in the trial. The mortality and frequency of pulmonary HMD on postmortem specimens were high in infants with RG and low in the remaining patients. Ninety-seven percent of the 31 patients who died with HMD proved at autopsy had RG on chest films. The mortality rate was lower in the treated group than in the controls. The combined effect of lower mortality and longer survival of fatal cases was such that the survival curve of treated patients was significantly different from controls. The treatment was studied in infants divided into subgroups according to sex, birth-weight, respiratory rate, blood pressure, and blood pH on admission. Mortality was lower in subgroups of treated patients than in control subgroups. There was a beneficial effect in the treated patients with low pH on admission and in those with severe hypotension as shown by the survival of the Ss. In babies who died, a transient increase of the average systolic blood

pressure was noted after the onset of treatment, whereas in controls a further decrease occurred. (33 refs.) - F. J. McNulty.

Department of Pediatrics
University of Rome
Rome, Italy

- 2287 ASNES, RUSSELL S., & LAMB, JOHNNY, M.
Neonatal respiratory depression secondary to maternal analgesics, treated by exchange transfusion. *Pediatrics*, 43(1):94-96, 1969.

Approximately 2 hours after birth, an apparently normal infant developed respiratory depression which was thought to be secondary to drugs administered during labor; he subsequently responded well to a double volume exchange transfusion. The implicated drug was apparently glutethimide (or possibly propiomazine). Idiosyncratic sensitivity and/or immaturity of the newborn may result in respiratory depression from these generally safe drugs. The differential diagnosis of respiratory depression should include drug toxicity and the treatment of choice is to remove the offending agent from the system. (7 refs.) E. L. Rowan.

601 North Broadway
Baltimore, Maryland 21205

- 2288 DAILY, WILLIAM J. R., *KLAUS, MARSHALL, & MEYER, H. BELTON P. Apnea in premature infants: Monitoring, incidence, heart rate changes, and an effect of environmental temperature. *Pediatrics*, 43(4):510-518, 1969.

Twenty-two premature infants (mean birth-weight of 1,417 gm) were continuously monitored for heart rate and apnea, and it was concluded that such monitoring is feasible and desirable. The respiratory pattern of the infants was continuously monitored by means of changes in electrical resistance across the chest during breathing (impedance plethymography). Seven of the infants were recovering from respiratory distress syndrome, one had pneumonia, and 14 were clinically well. A total of 540 apneic episodes (lasting more than 20 sec) occurred in 13 of the 22 infants with a daily incidence of apnea of 25% of the infants being studied that day. Bradycardia (heart rate less than 100 beats/min) was associated with the apnea and was always present after 30 seconds duration. Babies continued to be pink and have good tone for the first 20-30 seconds but cyanosis, pallor, and loss of muscle tone rapidly appeared

after 30 seconds. In 6 separate premature infants, it was found that apnea was more frequent at 36.8°C than at 36.0°C. It is suggested that monitoring of premature infants will detect apnea and prevent the sequence of cyanosis, bradycardia, and hypotonia. This may reduce the incidence of brain damage in premature infants. (26 refs.) - W. J. Klein.

Babies and Children's Hospital
Cleveland, Ohio

- 2289 FETUS AND NEWBORN COMMITTEE. CANADIAN PEDIATRIC SOCIETY. Oxygen therapy in the pre-term infant. *Canadian Medical Association Journal*, 99(11):564-567, 1968.

Newborn infants should be given an oxygen concentration that will produce blood oxygen tensions high enough to support life and avoid the dangers of hypoxemia, but low enough to avoid the danger of retrolental fibroplasia from hyperoxemia. Principles recommended for oxygen therapy in newborn infants are: the inspired oxygen concentration should be 10% higher than that required to relieve the cyanosis; arterial blood oxygen measurements should be used; and the oxygen administration should be stopped as soon as the improvement in the infant's condition permits. An oxygen concentration that is adequate for an infant while he is breathing should not be increased simply to avoid the cyanosis which may result from a neglected apneic episode. An apneic episode can be terminated with an auditory or tactile stimulus or a few puffs of ventilation by a face mask. There are no clinical signs for detecting hyperoxia, so if an infant is to undergo several days of oxygen therapy, he should be placed in a center where arterial blood gas measurements can be made. An oxygen analyzer should be used to determine the inspired oxygen and used to guide the oxygen flow adjustments. The oxygen analyzer should be checked at least once daily against room air and 100% oxygen to make sure the reading is within 2% true concentration. Ophthalmoscopic examination should be performed on infants under 2,000 gm birth-weight who have received oxygen therapy. (No refs.) - F. J. McNulty.

Canadian Pediatric Society
14 Green Avenue
St. Lambert, Quebec, Canada

- 2290 Sulfur compound may be breath of life for some babies. *Medical World News*, 10(16):G16, 1969.

Weak newborns, prematures, twins, and babies of diabetic mothers may benefit from an artificial atmosphere of O_2 and sulfur hexafluoride which will prevent hyaline membrane disease. In this disorder, the lungs can absorb too much oxygen and develop a protein membrane, pulmonary edema, congestion, and lung collapse. A newly devised artificial air with large sulfur molecules will partially block the oxygen from being absorbed. The artificial atmosphere has been tested only on animals. (No refs.) - M. Plessinger.

- 2291 VAN PRAAGH, IAN G. L., & TOVELL, HAROLD M. M. Cesarean section for fetal distress. *Obstetrics and Gynecology*, 31(5):674-681, 1968.

Fetal distress as measured by fetal heart rate alterations and changes in the amniotic fluid was studied by examining the charts of women delivered by cesarean section during the period between 1958 and 1966. For 192 of the cesarean sections, fetal distress was indicated, and there were 194 neonates who were classified according to the one-minute Apgar score (35 were severely depressed; 56 had scores of 4-6; 103 had scores of 7-10). The mothers' ages were within the normal range, and low segment cesarean section was performed 185 times, 10 of which were repeats. Forty-three patients underwent no labor; in 7, labor was induced; and 135 mothers received medication. Of the perinatal group, 55% weighed less than 2,500 gm. The membranes were intact at the time of cesarean section in 41% of the patients. Of the low Apgar group, 63% of the perinatal deaths and 50% of the survivors had stained amniotic fluid as compared to 47% and 36% of the other Apgar groups. Fetal heart bradycardia of less than 110/minute appeared in 50% of the 1-3 Apgar score groups. The most prominent signs of fetal distress were the combination of thick meconium and bradycardia (as determined in 62% of the perinatal deaths, 35% of the 1-3 Apgar scores, 14% in the 4-6 Apgar group, and 10% in the 7-10 Apgar group). Least significant was fetal irregularity. Molding of the vertex during labor which produces neural reflexes and pressure could alter the fetal heart rate. Sustained fetal tachycardia may be the earliest sign of fetal distress. (25 refs.) - V. G. Votano.

Women's Hospital
Saint Luke's Hospital Center
New York, New York 10025

- 2292 STENGER, VINCENT G., BLECHNER, JACK N., & PRYSTOWSKY, HARRY. A study of prolongation of obstetric anesthesia. *American Journal of Obstetrics and Gynecology*, 103(7):901-907, 1969.

Clinical and biochemical effects of prolonging obstetric anesthesia with pentothal, nitrous oxide (N_2O), and succinylcholine during cesarean section were studied in 13 normal pregnant Ss and their born and unborn offspring. Maternal respiratory gases, acid-base levels, and uterine metabolism did not vary greatly. The N_2O level in Ss with prolonged anesthesia was significantly higher in the uterine vein, fetal and neonate blood; the depressed clinical condition of the neonate was probably caused by N_2O narcosis, although the pentothal cannot be excluded. With the high N_2O blood level, blood oxygenation was effectively delayed; therefore, narcosis of the neonate is a primary complication of prolonged anesthesia. Pentothal, N_2O , and succinylcholine given for short times are an excellent anesthesia for cesarean sections. (7 refs.) - R. K. Butler.

University of Florida College of Medicine
Gainesville, Florida 32603

- 2293 LINTS, CARLTON E., & HARVEY, JOHN A. Drug induced reversal of brain damage in the rat. *Physiology and Behavior*, 4(1):29-31, 1969.

Male albino rats 90-95 days old received either sham-operations or bilateral lesions produced electrolytically in the medial fore-brain bundle, and 16 days later, the rats were tested for sensitivity to electric-foot shock. The lesions produced a 47% decrease in the telencephalic serotonin content and a 41% decrease in the jump threshold of the experimental animals. Intraperitoneal administration of DL-5-hydroxytryptophan 30 minutes before the test session had no effect on the jump threshold of the sham-operated animals, but increased that of the experimental rats in a dose-dependent manner, with a dose of 75 mg/kg causing complete reversal. Sensitivity of the lesioned rats was not affected by administration of D-5-hydroxytryptophan or L-DOPA, but was completely reversed with a dose of 37.5 mg/kg L-5-hydroxytryptophan. This reversal is probably due to increased endogenous serotonin, a decarboxylation product of L-5-hydroxytryptophan. (10 refs.) M. G. Conant.

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The University of Chicago
Chicago, Illinois 60637

2294 ALTCHER, ALBERT, ALBRIGHT, NILE L., & SOMMERS, SHELDON C. The renal pathology of toxemia of pregnancy. *Obstetrics and Gynecology*, 31(5):595-607, 1968.

Renal biopsies were performed in 76 women with a clinical diagnosis of toxemia of pregnancy (edema, hypertension, and proteinuria). Light microscopic examination revealed the pathognomonic picture of a glomerular lesion, juxtaglomerular cell hyperplasia, local epithelial degeneration of the loop of Henle, and afferent arteriolar spasm. Glomeruli were enlarged and cellular with hyperplastic intercapillary (mesangial) cells, swollen capillary endothelial cells, and fibrillar protein strands in Bowman's space. Most lesions cleared as clinical toxemia disappeared, but some residual increase in mesangial matrix was observed. Biopsy rather than autopsy examination is necessary if one seeks to understand the mechanism of a pathophysiological disease. (56 refs.) - E. L. Rowan.

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2295 Brain lesions from blunt trauma in early infancy. *Journal of the American Medical Association*, 207(11):2094-2095, 1969. (Editorial)

Traumatic lesions in brains of infants are characterized by tears in the white matter in infants under 5 months of age; while in older infants, the traumatic lesions are the same as those in the adult brain. There are generally some clinical repercussions in the tissue loss from tears in such brain-injured patients, and this tissue loss may result in defective mental, neurological, and speech development. (3 refs.) - S. Half.

2296 PEYSER, M. R., & TOAFF, R. Traumatic rupture of the placenta: A rare cause of fetal death. *Obstetrics and Gynecology*, 34(4):561-563, 1969.

Traumatic rupture of placenta during the thirty-fifth week of pregnancy was caused by an automobile accident. Delivery of a macerated fetus occurred 5 days later. The patient had no injuries apart from the bruises of her knees; however, the mechanism of the placental rupture may be understood by reconstructing the accident. During the accident, the abdomen of the patient hit the thighs causing a sudden reduction of the antero-posterior diameters of the uterus. Because

the placenta lacked the elasticity of the uterus, it gave way at the point of attachment to the uterus. The bleeding from both maternal and fetal circulation was drained into the amniotic sac and thus caused the death of the fetus by exsanguination. (9 refs.) - L. S. Ho.

Maternity Hospital Hakirya
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Tel-Aviv, Israel

2297 KALVERBOER, A. F. A study of "minimal brain dysfunction." *Developmental Medicine and Child Neurology*, 11(1):115-116, 1969. (Letter)

Most studies of "minimal brain dysfunction" are filled with methodological pitfalls. In order to be reproducible, a study must rigidly define its purpose, basic concepts, and experimental design. The data collected must then be relevant to the purpose for which the study was undertaken. Until these criteria are met, little progress will be made in understanding the relationships between neurological condition and behavior. (1 ref.)
E. L. Rowan.

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Oostersingel 59
Groningen, The Netherlands

2298 STERNGLOSS, ERNEST J. Infant mortality and nuclear tests. *Bulletin of the Atomic Scientists*, 25(4):18-20, 1969.

There is a close correlation between fetal and infant death rates and the accumulation of strontium-90 on the ground and between the leveling off in the decline of death rates in the high rainfall areas of the East and South in the year 1951-1952 and the onset of the Nevada nuclear weapons tests in the atmosphere. The dry Western states showed a steady decline in fetal and infant death rates until 1954-1958 when stratospheric debris from the tests began to be introduced into the atmosphere. A similar situation existed after the tests in Nevada in 1954. It is to be expected that the first serious effects of radiation fallout would appear in the fetus and infant, where rapid cell division is extremely sensitive to

radiation. The excess fetal and infant deaths may be primarily due to chromosomal damage produced just prior to conception or in the earliest phases of development. (1 ref.)
M. G. Conant.

Department of Radiology
Division of Radiation Health
University of Pittsburgh
Pittsburgh, Pennsylvania 15213

2299 STERNGLASS, ERNEST J. Evidence for low-level radiation effects on the human embryo and fetus. In: Sikov, M. R. *Radiation Biology of the Fetal and Juvenile Mammal*. U. S. Atomic Energy Commission, the Technical Information Division, 1969, p. 693-717.

The childhood leukemia rate in the Troy-Albany, New York area doubled in the 8 years following the rainout of radioactive debris from a test blast in Nevada in April 1953. The increase was characterized by a 4-5 year delay in onset and a shift in age distribution at onset toward older age which indicates that low-level and low-dose-rate effects can occur prior to conception. Analysis of fetal, neonatal, and postnatal mortality rates reveals geographical changes in the rates which coincide with known long-range fallout patterns in the United States. Changes in the mortality rates are also closely correlated with the measured amounts of strontium-90 in the milk and in the bone and teeth of the fetus and newborn. The developing human embryo and fetus are highly sensitive to both acute radiation at diagnostic levels and to low dose-rate radiation from fallout, with the latter causing genetic damage prior to conception. The lack of increases in incidence of leukemia in Japan following the bombing indicates that acute radiation does not produce the genetic damage that low-level radiation does. (37 refs.)
M. G. Conant.

2300 PHEMISTER, ROBERT D., SHIVELY, JAMES N., & YOUNG, STUART. The effects of gamma irradiation on the postnatally developing canine cerebellar cortex. I. Effects of single sublethal exposures. *Journal of Neuropathology and Experimental Neurology*, 28(1): 119-127, 1969.

Morphologic effects were produced in the cerebellum of 30 beagle puppies by single doses of 85, 170, 325, or 500 R cobalt-60 γ radiation on the second postnatal day. The mean widths of both the external and internal granular layers were significantly decreased

by each of the 4 doses, the total cortical layer width by 3 exposures (170, 325, and 500 R), the mean width of the molecular layer by the 325 and 500 R dose levels, and the Purkinje cell layer by only the 500 R dose level. Marked histologic malformations, including ectopic clusters and poorly defined rows of granule cells in the molecular layer and of localized areas with reduced cell numbers in the internal granular layer, were observed following doses of either 325 or 500 R. Dogs that received doses of 85 or 170 R showed no such changes. (20 refs.) - M. G. Conant.

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Fort Collins, Colorado 80521

2301 PHEMISTER, ROBERT D., SHIVELY, JAMES N., & YOUNG, STUART. The effects of gamma irradiation on the postnatally developing canine cerebellar cortex. II. Sequential histogenesis of radiation-induced changes. *Journal of Neuropathology and Experimental Neurology*, 28(1):128-138, 1969.

Total-body exposure of 325 R cobalt-60 γ radiation administered on the second postnatal day to 48 beagle puppies caused pathological changes in the cerebellum on days 2, 10, 30, and 70 after irradiation. By the second post-irradiation day, massive selective destruction of the external granular layer had occurred, and though partial recovery followed, this was insufficient to prevent permanent thinning of the internal granular layer. The molecular layer grew normally until the thirtieth post-irradiation day when it began to slow down which suggests an effect on the production of fine dendritic branches of Purkinje cells. The reduced total cortical thickness was due, at first, to destruction of the external granular layer, but later in the post-irradiation period, to destruction of the internal granular layer and, by 70 days, the molecular layer. Ectopic granule cell formations were observed in the molecular layer by the thirtieth day after exposure, rosette formation was not observed, and effects on postnatal myelination were mild. (16 refs.) - M. G. Conant.

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- 2302 GUSTAVSON, K. H., * HAGSBERG, B., & SANNER, G. Identical syndromes of cerebral palsy in the same family. *Acta Paediatrica Scandinavica*, 58(4):330-340, 1969.

A survey of almost all known cases of cerebral palsy in Sweden revealed 43 families (98 cases) where more than one family member was affected. Of these, 16 families (43 cases) showed identical, non-progressive syndromes with a normal perinatal history, and these were important from a genetic point of view. Chromosomal studies and laboratory screening tests were normal in this group. A disproportionate number of these families had ataxic syndromes. Congenital ataxia (10 families) was associated with moderate to severe MR. The identical syndrome in siblings, both sexes affected, healthy parents and consanguinity (3 families) suggests an autosomal recessive mode of inheritance. Ataxic diplegia (3 families) was inherited as an autosomal dominant or autosomal recessive trait, and only the former was associated with normal intelligence. The spastic and dyskinetic syndromes which make up 75% to 90% of cases of cerebral palsy in Sweden were seen in only 3 families. Although heredity accounts for only a small proportion (1.5%) of cerebral palsy in Sweden, it is an etiologic factor which cannot be ignored. (43 refs.)

E. L. Rowan

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Akademiska Sjukhuset
Uppsala, Sweden

- 2303 SYLVESTER, P. E. Pyramidal, lemniscal and parietal lobe status in cerebral palsy. *Journal of Mental Deficiency Research*, 13(1):20-33, 1969.

The anatomical and pathological status of the cortico-spinal tract, somatosensory tracts,

and parietal lobes of the brains of 11 patients with cerebral palsy and MR was compared with that in 11 control brains. In spastic patients, the weight of the medullary pyramids, as well as the number of both large and small fibers within them, was significantly less than controls. Parietal lobe weights were also smaller in spastics than in controls; however, there was no difference in medial lemniscal size. Up to the level of the brainstem the somato-sensory tracts were apparently intact. Because of reduced cortical size, however, afferent fibers have less opportunity to register their information at that level. (14 refs.) - E. L. Rowan.

St. Lawrence's Hospital
Caterham, Surrey
England

- 2304 NATHAN, P. W. Treatment of spasticity with perineural injections of phenol. *Developmental Medicine and Child Neurology*, 11(3):384, 1969. (Annotation)

A number of nerve fibers are destroyed by the injection of phenol and alcohol solutions around nerve roots or peripheral nerves; however, this treatment of spasticity in CP children is not recommended because of the side-effects and the transient nature of the relief. (3 refs.) - S. Half.

National Hospital for Nervous Diseases
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London, W. C. 1, England

Diseases or disorders of metabolism, growth, or nutrition

- 2305 SCRIVER, CHARLES R. Inborn errors of amino-acid metabolism. *British Medical Bulletin*, 25(1):35-41, 1969.

New knowledge concerning hereditary amino-acidopathies will benefit the afflicted individual and mankind in general. Heretofore, exact phenotypic replicas of the human conditions were not duplicated in animals;

therefore, extensive study has been given to cultured tissue explants which are used for investigation of the mutant phenotype and for evaluation of membrane transport characteristics. Since recognition of abnormality depends on a definition of normal limits, the normal distribution of free amino acids in body-water and normal transport of free amino acids were studied in detail and it was found

that each amino acid utilizes a specific transport system. Mutant transport systems for each amino acid will probably be discovered in the future. Enzyme activity is also important, as many investigators feel that a spectrum of enzyme variation can be measured. Enzyme studies may also permit genotypic variants with common metabolic phenotypes to be separated from one another. The use of screening tests on infants permits the early detection of aminoacidopathies and the application of proper treatment. (110 refs.)

M. G. Conant.

McGill University-Montreal Children's Hospital
Montreal, Quebec, Canada

2306 Mass screening for aminoacid disorders.
Lancet, 1(7606):1133-1134, 1969.
(Editorial)

The possibility of early detection of inborn metabolic diseases has resulted from the mass screening of newborns for amino acid disturbances. The most prevalent inborn metabolic error is PKU. Urine should be tested when the infant is 4 weeks old, and blood should be tested between the ages of 5 and 10 days. Non-treated amino acid anomalies may cause neurological damage, but once the carriers are detected, eugenic counseling will help to reduce these diseases in the community. Additional techniques and further studies are indicated for the newborn. These biochemical disorders should be studied intensively to detect other disorders and to find an effective regimen of treatment. (13 refs.)

S. Half.

2307 NYHAN, WILLIAM L. Genetic defects of amino acid metabolism. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 14, p. 868-895.

Phenylketonuria and homocystinuria are the most frequently encountered hereditary disorders of amino acid metabolism among MR patients. Dietary restriction of phenylalanine improves eczema, behavior, and neurologic and EEG pictures in phenylketonuria. MR is also found in histinemia, hypersarcosinemia, hyperprolinemia, hydroxyprolinemia, cystathioninuria, methioninemia, argininosuccinic aciduria, citrullinemia, hyperammonemia, hyperlysinemia, and Hartnup disease. Tyrosinosis, alkaptonuria, albinism, imidazole aminoaciduria, maple syrup urine disease,

hypervalinemia, hyperglycinemia, hyperoxaluria and oxalosis, and cystinuria are necessarily not associated with MR. Where genetic data are available, these disorders all seem to be transmitted as an autosomal recessive trait. (166 refs.) - L. S. Ho.

2308 CHURCHILL, J. A., *MOGHISSI, K. S., EVANS, T. N., & FROHMAN, C. Relationships of maternal amino acid blood levels to fetal development. *Obstetrics and Gynecology*, 33(4):492-495, 1969.

The total α -amino acid concentration was determined in blood samples from 64 pregnant Negro women (sample taken at 32-34 and 34-36 wks of gestation) and averaged (mean value of 3.992 mg/100 ml). The maternal α -amino acid concentration was directly related to the birth-weight of the baby and to its cranial volume; however, there was no clear relation between estimated dietary intake and the status of the infant nor maternal amino acid values. Fetal development may be stunted when amino acids available to the fetus are diminished, and high maternal blood amino acid levels during pregnancy may be important in reducing the incidence of low birth-weight babies. (8 refs.) - M. G. Conant.

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2309 GOLDSTEIN, NORMAN P., EWERT, JOSEPHINE C., RANDALL, RAYMOND V., & GROSS, JOHN B. Psychiatric aspects of Wilson's disease (hepatolenticular degeneration): Results of psychometric tests during long-term therapy. *American Journal of Psychiatry*, 124(11):1555-1561, 1968.

In order to verify the observed association between mental impairment and Wilson's disease, a group of 22 patients with hepatolenticular degeneration was evaluated psychiatrically and followed psychometrically during the course of treatment. Nine of these patients were considered to be psychiatrically ill, although there was no particular pattern of personality disturbance. One boy was considered to be both MR and schizophrenic. During the course of therapy with penicillamine, and low copper diet, there was a gradual improvement in psychological test scores for the 13 patients so tested. Again there were no specific patterns, but there was general improvement in comprehension, concept formation, and recall. Some individuals showed dramatic improvement in measured intelligence

and others in reduced anxiety. The MR boy deteriorated psychologically despite neurologic improvement. With continuous treatment of Wilson's disease, one may expect gradual improvement in psychological test scores; however, the reasons for such improvement and the possible role of copper deposition in cortical function are still unclear. (3 refs.) - E. L. Rowan.

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2310 SCHWARTZ, JAMES F., & KOLENDARIANOS, ERNEST T. Maple syrup urine disease: A review with a report of an additional case. *Developmental Medicine and Child Neurology*, 11(4):460-470, 1969.

A case of maple syrup urine disease (MSUD) is presented, with a discussion of the characteristic clinical syndrome of neonatal feeding difficulty, lethargy, respiratory irregularity, seizures, and opisthotonos. The biochemical abnormality in the oxidative decarboxylation of the keto-acids of valine, leucine and isoleucine is described, and an approach to dietary therapy, by carefully controlling the amino-acid composition of the diet, is outlined. The pathological findings are widespread, but with especially marked alterations in myelin formation in the cerebral hemispheres. This unusual disorder, which is one of the classic inborn errors of metabolism, is no longer merely of academic importance. With the development of diet therapy and means of controlling the metabolic derangement, thereby preventing mental retardation and widespread neurological deficits, the diagnosis of MSUD and early initiation of therapy becomes an urgent and intensely practical matter, especially in the immediate neonatal period. (29 refs.)
Journal summary.

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2311 DICKINSON, J. P., HOLTON, J. B., LEWIS, G. M., LITTLEWOOD, J. M., & STEEL, A. E. Maple syrup urine disease: Four years' experience with dietary treatment of a case. *Acta Paediatrica Scandinavica*, 58(4):341-351, 1969.

An infant with maple syrup urine disease (MSUD) presented with a typical picture of failure-to-feed, signs of intracranial disturbance, and high systemic and urinary concentrations of the branched-chain amino acids

(leucine, isoleucine, and valine) and was begun on dietary treatment at 6 weeks of age. A commercial preparation without branched-chain amino acids was given initially and supplemented by vitamins and essential quantities of these amino acids as indicated by frequent plasma amino acid measurement. Despite this careful management and subsequently good physical development, the child functioned at an 18-month-old MA when 4 1/2 years of age. Review of the 55 reported cases of MSUD revealed that 28 of the 30 who received no dietary treatment were dead and the 2 survivors were MR. Eight of the 25 treated cases were alive but only 4 were mentally normal. In these 4, treatment had begun before the tenth day of life. A child with MSUD is salvageable, but early diagnosis and treatment are mandatory and constant monitoring of plasma amino acids is essential since even a mild stress (infection) can upset delicate protein balance and cause central nervous system damage. (75 refs.)
E. L. Rowan.

Department of Pathology
School of Medicine
Leeds 2, England

2312 GOEDDE, H. WERNER, LANGENBECK, ULRICH, & BRACKERTZ, DIETER. Detection of heterozygotes in maple syrup urine disease: Role of lymphocyte count. *Humangenetik*, 6(2):189-190, 1968.

Determination of enzyme activities for the decarboxylation of branched chain α -keto acids in a leukocyte suspension showed that a linear relation exists between the CO_2 produced from the α -keto acids and the relative lymphocyte count for substrates 1- ^{14}C -labeled α -ketoisocaproic acid, α -ketoisovaleric acid, and α -keto- β -methylvaleric acid. Equations to represent the dependence of enzyme activity and lymphocyte contents are $y=0.17+0.0584Nx$ for α -ketoisocaproic acid (where N =number Ss tested and x =lymphocyte content in percent of 16×10^6 leukocytes tested); $y=0.73+0.0528Nx$ for α -ketoisovaleric acid; and $y=0.48+0.0313Nx$ for α -keto- β -methylvaleric acid. The May-Grünwald-Giemsa method of staining smears was used to determine the lymphocyte content of the leukocyte suspension for 23 normal homozygotes. (8 refs.) - A. Huffer.

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- 2313 McMILLAN, MARY. Screening for phenylketonuria. *British Medical Journal*, 1(5638):252, 1969. (Letter)

The implication that the Report of the Medical Research Council Working Party on Phenylketonuria (PKU) recommends the Guthrie test on blood as the preferred test to replace the Phenistix test in screening for PKU is not justified. The plasma chromatographic test is as easy and inexpensive as the Guthrie blood test and has the added advantage of detecting other inborn metabolic errors. (2 refs.) - E. F. MacGregor.

Lewisham Group Laboratory
London S.E. 13, England

- 2314 WARWICK, WARREN J. Phenylketonuria and the practice of medicine. *Journal of the American Medical Association*, 207(11):2095, 1969.

The success and methodology used in early detection of PKU in newborn patients can serve as a model to physicians interested in establishing the practice of preventive medicine. The experience of the PKU Study Center can assist young physicians in developing clinical therapy specialties and special centers for experimental approaches of a therapeutic, preventative nature. Early diagnosis and treatment of diseases are imperative as is the establishment of mutual confidence in the doctor-patient contract. (No refs.)

S. Half.

No address

- 2315 CUNNINGHAM, GEORGE C., DAY, ROBERT W., BERMAN, JULIAN L., & HSIA, DAVID YI-YUNG. Phenylalanine tolerance tests. *American Journal of Diseases of Children*, 117(6):626-635, 1969.

Families with phenylketonuria (PKU) and hyperphenylalaninemia (HPA) were administered phenylalanine tolerance tests, and the mean serum phenylalanine and tyrosine values after selected loading in affected siblings with HPA were lower than those in affected siblings with PKU. The values for the unaffected siblings of HPAs were not different from unaffected siblings and parents of PKUs. Phenylalanine tolerance tests were performed on index patients, their siblings, and their parents in 122 families and in control groups of comparable age and sex. The families were divided into group 1 with a blood phenylalanine of 20 mg/100 ml or higher and group 2

patients with HPA and a blood phenylalanine level less than 19.9 mg/100 ml. Group 1 was further subdivided into group 1A or typical PKU, where at least one older sibling of the proband was affected and had MR; group 1B or atypical PKU where an affected sibling had normal mental development, and group 1C in which the proband was the only one affected in the sibship or in which patients were unclassified by these criteria. The reproducibility for phenylalanine tolerance expressed as coefficient of variation was 7%. There was no difference in the tolerance of unaffected siblings from group 1 with an IQ 84 or more and a group with lower IQs. Previous phenylalanine intake has little effect in determining loading response. The persistent elevation of phenylalanine may be due to the existence of a modifier gene. (33 refs.)

F. J. McNulty.

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Bureau of Maternal and Child Health
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- 2316 HILL, JOHN B. A climatological factor influencing the determination of phenylalanine in blood of newborn infants in North Carolina. *Biochemical Medicine*, 2(4):261-273, 1969.

The determination of blood phenylalanine levels by the punch disc technique was found to be related to variations in relative humidity. In 1965, the state of North Carolina began a screening program for phenylketonuria, and from July 1965 through February 1968, the blood phenylalanine showed seasonal changes in level in which a lower level was reached in July or August, and an upward trend, which continued through April, then emerged. The monthly mean phenylalanine blood level data obtained during the first 8 months of screening was plotted against the relative humidity recorded at the Raleigh-Durham weather station for the same 8 months. A high degree of inverse correlation was noted which indicated the screening program using the punched discs for phenylalanine values was subject to the influence of relative humidity. Experiments to determine the amount of effect of humidity in the punched disc technique found that samples kept at 100% relative humidity contained significantly less blood urea nitrogen in the punched spot. If a degree of precision is needed, then known volumes of capillary blood should be placed on the filter paper and eluted in their entirety; however, for the screening of large populations, the filter

punching technique allows inexperienced personnel to accurately measure small volumes and to obtain blood samples efficiently. (11 refs.) - F. J. McNulty.

University of North Carolina School
of Medicine
Chapel Hill, North Carolina 27514

2317 EFRON, MARY L., KANG, ELLEN SONG, VISAKORPI, JARMO, & FELLERS, FRANCIS X. Effect of elevated plasma phenylalanine levels on other amino acids in phenylketonuric and normal subjects. *Journal of Pediatrics*, 74 (3):399-405, 1969.

Many plasma amino acids of 20 untreated PKUs were significantly lower than those of 20 matched MR, non-phenylketonuric patients and 10 normal control Ss. Two Ss with phenylketonuria and one normal S responded similarly to a phenylalanine infusion by a lowering of plasma essential and nonessential amino acids. Renal tubular resorption of amino acids was not significantly changed by phenylalanine infusion in these 3 Ss. The plasma amino acid concentrations were not significantly different between a relatively intelligent patient with PKU and her sib with a very low IQ, with the exception of citrulline and half cystine. Low plasma amino acid concentrations may reflect low intracellular amino acids which in turn may result in poor synthesis of brain proteolipids and other proteins necessary for normal mental development. (17 refs.) - L. S. Ho.

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2318 COHEN, B. E., SZEINBERG, A., AVIAD, Y., & COSTEFF, H. Evaluation of dietary treatment in phenylketonuria: A proposed methodology. *Developmental Medicine and Child Neurology*, 11(1):96-102, 1969.

Newborn infants with phenylketonuria (PKU) were ranked by adequacy of dietary control, IQ, and age of starting treatment with normal sibling controls; the findings indicated that a well-controlled early diet probably is significantly associated with higher IQs. The 10 treated patients have a mean IQ of 61 compared with a mean IQ of 46 (not significantly different) in the 6 untreated patients. The 8 patients treated from 3 months of age or earlier have a mean IQ of 68 and the 4 early-treated patients, with at least 70% of all blood phenylalanine levels below 10 mg/100 ml, have a mean IQ of 76. The method uses the

difference between the IQ of the patient and of the normal sibling control which should enable international data to be pooled. Only cases determined by screening newborns or screening siblings of previously diagnosed PKU cases are used in this type of analysis. The cases analyzed are too few for a satisfactory weighing of the relative values of chemical control and age of beginning diet in the final treatment results. The analysis of pooled results of PKU treatment are dependent on the method by which PKU was determined (population screened to find the cases; age of onset and duration of treatment; IQ and age at testing; index of adequacy of chemical control; and IQ of an age-matched normal sibling control). (8 refs.) - F. J. McNulty.

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Hospital
University Medical School
Tel-Hashomer, Israel

2319 BAGCHI, S. P., & ZARYCKI, EDWIN P. In vivo formation of tyrosine from phenylalanine in brain. *Life Sciences*, 9(Part I): 111-119, 1970.

Attempts to demonstrate phenylalanine hydroxylase, which converts phenylalanine to tyrosine, have not succeeded, but tyrosine hydroxylase, which can convert phenylalanine to tyrosine and the latter into 3,4-dihydroxyphenylalanine (DOPA), has been demonstrated *in vitro*. Therefore, the possibility that phenylalanine is hydroxylated to tyrosine by rat brain *in vivo* was examined. Twenty minutes after the administration of phenylalanine-¹⁴C into lateral brain ventricles, radioactivity was incorporated into brainstem and cerebellar tyrosine. Small, but significant, amounts of radioactivity were also incorporated into brainstem norepinephrine. The intraperitoneal administration of *p*-chlorophenylalanine, which abolishes the action of hepatic phenylalanine hydroxylase, did not inhibit label incorporation into brain tyrosine. Thus, the slight conversion of phenylalanine into tyrosine seen in patients with phenylketonuria probably is due to the presence of central nervous system tyrosine hydroxylase. (17 refs.) - K. Jarka.

Psychiatric Research Unit
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Saskatoon, Saskatchewan, Canada

2320 MCKEAN, C.M., BOGGS, D.E., & PETERSON, N. A. The influence of high phenylalanine and tyrosine on the concentrations of essential amino acids in brain. *Journal of Neurochemistry*, 15(2):235-241, 1968.

Male rats were subjected to high concentrations of phenylalanine or tyrosine, either by intraperitoneal injection or as a 7% dietary supplement, and the concentrations of the essential amino acids in the brain were determined 15, 30, or 60 minutes after injection or after 3 weeks on the experimental diet. High circulating levels of phenylalanine caused depletions of threonine, valine, methionine, isoleucine, leucine, histidine, tryptophan, and tyrosine, with the branched-chain amino acids most affected (their concentrations were reduced to 38-64% of the control values). Similar cerebral patterns were found in both adult and infant rats, while phenylalanine loading produced depletions in serum amino acid levels in adult rats, but unchanged or slightly elevated levels in infant rats. Similar changes, but on a smaller scale, were observed following tyrosine loading. High blood phenylalanine levels reduced the brain:blood ratio of most of the essential amino acids in both age groups. (9 refs.) - M. G. Conant.

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Eldridge, California 95431

2321 GENTZ, J., HEINRICH, J., *LINDBLAD, B., LINDSTEDT, S., & ZETTERSTROM, R. Enzymatic studies in a case of hereditary tyrosinemia with hepatoma. *Acta Paediatrica Scandinavica*, 58(4):393-396, 1969.

A 16-year-old girl with typical hereditary tyrosinemia and a hepatoma had an elevated urinary excretion of Δ -aminolevulinic acid, a finding noted in 6 other patients with hereditary tyrosinemia. Activity of Δ -aminolevulinic acid synthetase was 7 times higher in hepatoma tissue than in surrounding liver tissue where it approximated control values. Tyrosinemia results from an inability to form homogentisate from *p*-hydroxyphenylpyruvate, and the enzymes of tyrosine metabolism (*p*-hydroxyphenylpyruvate hydroxylase and tyrosine aminotransferase) were absent in hepatoma tissue and less than normal in surrounding tissue. The altered activity of these 2 enzyme systems may be coincidental, but a biochemical relationship between these metabolic pathways is suggested. (22 refs.)

E. L. Rowan.

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22007 Lund 7, Sweden

2322 LABERGE, CLAUDE. Hereditary tyrosinemia in a French Canadian isolate. *American Journal of Human Genetics*, 21(1):36-45, 1969.

Among the isolated population of the Lake St. John-Chicoutimi area of Quebec, Canada are 29 families who have had 39 children with hereditary tyrosinemia manifest as acute hepatic failure of infancy. Pedigree study has traced all affected children to a single ancestral pair, at least one of whom brought the gene from France to Canada in 1644. The disorder is inherited as a mendelian autosomal recessive trait. Despite the fact that there were no first-cousin marriages in the families studied, the high carrier rate of 1:24 to 1:41 in the isolated population enhances the risks of affliction. (23 refs.)

E. L. Rowan.

Hopital St. Michel-Archange
Quebec (5), Canada

2323 TADA, KEIYA, WADA, YOSHIRO, YAZAKI, NATSUME, YOKOYAMA, YOSHIMASA, NAKAGAWA, HIROSHI, YOSHIDA, TOSHIO, SATO, TETSURO, & *ARAKAWA, TSUNEO. Dietary treatment of infantile tyrosinemia. *Tohoku Journal of Experimental Medicine*, 95(4):337-344, 1968.

Tyrosinemia was diagnosed in a 4-month-old male with developmental retardation, abdominal distension, and jaundice; a diet low in tyrosine (13 mg/kg) and phenylalanine (18 mg/kg) was instituted at 5 months of age. With this regimen, the child had no weight gain, and he developed an increased susceptibility to infection. Therefore, the diet was modified to provide 30 mg/kg tyrosine and 35 mg/kg phenylalanine daily. After 10 months of dietary restriction, there was nearly a complete disappearance of clinical symptoms including failure to thrive and renal and hepatic damage. At 15 months of age, the patient is of normal size with a somewhat retarded mental development. (10 refs.) - M. G. Conant.

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Sendai, Japan

2324 LEVY, HARVEY L., SHIH, VIVIAN E., MADIGAN, PHYLLIS M., & MacCREADY, ROBERT A. Transient tyrosinemia in full-term infants. *Journal of the American Medical Association*, 209(2):249-250, 1969.

Transient tyrosinemia has been found in approximately 1.8% of all infants in Massachusetts tested within the first 6 weeks of life

(2,085 infants out of a total screened population of 117,752). About 24% have been full-term by weight and gestational criteria. In general, infants with lower birth-weights had more marked tyrosinemia. However, several babies with average or above average birth-weights had blood tyrosine concentrations of 10 mg/100 ml to more than 20 mg/100 ml, and in a few of these latter infants, the tyrosinemia lasted as long as 9 to 13 weeks after birth. The infants have been clinically well. (9 refs.) - *Journal summary*.

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- 2325 VELLAN, E. J., GJESSING, L. R., & SEIP, M. Hair amino acids in cystinosis, homocystinuria, Folling's disease and tyrosinosis. *Acta Paediatrica Scandinavica*, 58(3): 287-289, 1969.

Alanine, cystine, homocystine, methionine, phenylalanine, and tyrosine levels were not significantly different from normal in the hair of patients with cystinosis, homocystinuria, tyrosinosis, or Folling's disease. It would appear, therefore, that the amino acid content in keratin will not be very useful in the differential diagnosis of amino acid inborn metabolic disorders nor in the detection of possible heterozygotes. (6 refs.) - *L. S. Ho*.

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Asker, Norway

- 2326 CUSWORTH, D. C., & DENT, C. E. Homocystinuria. *British Medical Bulletin*, 25 (1):42-47, 1969.

Homocystinuria is the next most common inborn error of metabolism, after PKU, in producing mental deficiency; the disorder is due to a block in the metabolic pathway between methionine and cystine. The clinically characteristic features include long limbs, dislocated lenses, glaucoma, MR, cardiovascular disorders, and epilepsy (some severe cases). The disease is due to an autosomal recessive gene. Patients show frequent thromboses in the veins and arteries, and platelet stickiness is found to be abnormally high. Therapeutic attempts have been directed toward the reduction of methionine and homocystine accumulated within the body and toward replacement of cystathionine which is deficient. The dietary treatment is difficult to manage, and full assessment will require many more years of experience; however, administration of

pyridoxine hydrochloride reduces methionine and homocystine levels with no adverse reaction in patients with varying severity of disease. (36 refs.) - *M. G. Conant*.

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- 2327 HAGBERG, BENGT, HAMBRAEUS, LEIF, & HAMFELT, ARNE. Pyridoxine in homocystinuria. *Lancet*, 2(7614):27, 1969. (Letter)

A 17-year-old boy who gradually developed the extrapyramidal symptoms of homocystinuria was treated with pyridoxine (300 mg daily) which provided temporary clinical improvement. His condition started to regress after 2 weeks although the plasma pyridoxal-5-phosphate level increased significantly the first day of treatment (from 6 mg/ml to 79 mg/ml) and remained high after 35 days (168 mg/ml). Since a similar situation occurred in the S's 2 siblings, it appears that improvement in plasma pyridoxal-5-phosphate levels is not necessarily accompanied by lasting improvement in the clinical course. (7 refs.) - *A. Huffer*.

Department of Pediatrics
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Uppsala, Sweden

- 2328 McCULLY, KILMER S. Importance of homocysteine-induced abnormalities of proteoglycan structure in arteriosclerosis. *American Journal of Pathology*, 59(1):181-193, 1970.

A deficiency of cystathionine synthetase is associated with MR and connective tissue disorders in humans. Cells cultured from the skin of humans with cystathionine synthetase deficiency synthesized abnormal proteoglycans that were granular, aggregated, and flocculent; this abnormality produced marked distortion of the normal fibrillar structure of the cells. Addition of *DL*-homocysteine (1 μ mole/ml) to cultures of normal skin cells induced the synthesis of abnormal proteoglycans and produced morphological changes similar to those seen in enzyme-deficient cell cultures. Analyses of the protein of cultured, normal cell monolayers showed that only traces of hydroxyproline are synthesized by normal cells and that the amino acid composition of these cells resembles that of connective tissue proteoglycans. This suggested that homocysteinemia may cause the synthesis of abnormal proteoglycans by blood-vessel cells in arteriosclerosis; therapy

which decreases endogenous and exogenous concentrations of homocysteine in such cells may be useful in the treatment of arteriosclerosis. (30 refs.) - K. Jarka.

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2329 WADA, YOSHIRO, *ARAKAWA, TSUNEO, & KOI-ZUMI, KINJIRO. Lesch-Nyhan syndrome: Autopsy findings and in vitro study of incorporation of ^{14}C -8-inosine into uric acid, guanosine-monophosphate and adenosine-monophosphate in the liver. *Tohoku Journal of Experimental Medicine*, 95(3):253-260, 1968.

Liver specimens obtained surgically from a 21-month-old male patient with Lesch-Nyhan syndrome and from a control patient without hepatic involvement were incubated with ^{14}C -8-inosine. The incorporation of ^{14}C -8-inosine into uric acid and xanthine was markedly increased, while that into guanosine-monophosphate, adenine, and adenosine was markedly decreased in the patient with Lesch-Nyhan disease. These findings support the theory that the primary defect in this syndrome is in the activity of hypoxanthine-guanine-phosphoribosyl transferase. At autopsy, the cerebrum had changed into a thin layer of gliosis containing nerve cells with degenerative and necrotic changes, and foci of necrosis of nerve cells were also noted in the cerebrum. Diffuse fibrosis was observed in the medulla of the kidney, and striking congestion was noted in the spleen. (7 refs.) - M. G. Conant.

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Sendai, Japan

2330 SHIH, VIVIAN E., EFRON, MARY L., & MOSER, HUGO W. Hyperornithinemia, hyperammonemia, and homocitrullinuria: A new disorder of amino acid metabolism associated with myoclonic seizures and mental retardation. *American Journal of Diseases of Children*, 117(1):83-92, 1969.

A new syndrome in a male child (CA 3 yrs), in which intermittent hyperammonemia was associated with abnormally high levels of ornithine in the blood and of homocitrulline in the urine, was accompanied by CNS disorders and developmental retardation. At the age of 16.5 months, the patient showed MR, irritability, myoclonic spasms, psychomotor retardation, and intermittent ataxia which

improved upon institution of a low-protein diet (1.5 g/kg/day) at 17.5 months. Dietary treatment also lowered blood ammonia and ornithine levels and resulted in clinical improvement. Other liver functions were only mildly abnormal, the parents were normal, lysine loading in the patient did not produce nor aggravate abnormalities, and slow disposal of ornithine followed oral loading. These results suggest that the disorder is due to a block in the ornithine metabolic pathway unlike that in patients with hepatic ornithine transcarbamylase deficiency. (22 refs.) - M. G. Conant.

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2331 LEVIN, B., OBERHOLZER, V. G., & SINCLAIR, L. Biochemical investigations of hyperammonaemia. *Lancet*, 2(7613):170-174, 1969.

Three cases of hyperammonemia, an inborn error of metabolism caused by severe hepatic ornithine-transcarbamylase deficiency, were investigated biochemically. The impairment of the urea cycle results in accumulation of ammonia in both blood and cerebrospinal fluid (CSF) which in turn leads to high levels of glutamine. It also causes an increased activity in pyrimidine metabolism leading to high urinary excretion of orotic acid, uridine, and uracil. Urea formation still continues, sufficient to maintain a low-normal blood-urea. It could be increased as measured by the rise in blood-level and urinary excretion when the protein intake is increased from a very low level to moderate ones. The blood-ammonia is most effectively reduced by lowering the protein intake. (19 refs.) - *Journal summary*.

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2332 STEIN, ISRAEL M., *COHEN, BURTON D., & KORNHAUSER, ROGER S. Guanidino-succinic acid in renal failure, experimental azotemia and inborn errors of the urea cycle. *New England Journal of Medicine*, 280(17):926-930, 1969.

An improved method of isolation and quantitation, using Dowex-1 ion-exchange chromatography and paper electrophoresis, has permitted efficient recovery of guanidinosuccinic acid from serum, urine, and cerebrospinal fluid. Mean concentration of this acid in normal urine is 1.71 ± 1.60 (SD) mg/100 ml, and 5.35 ± 2.68 mg/100 ml in urine

from uremic patients. Serum and cerebrospinal fluid levels are also increased in uremia. In patients undergoing peritoneal dialysis, guanidosuccinic acid was dialyzable. The metabolic pathway for synthesis of guanidosuccinic acid is unknown. The increased excretion of this compound in rats treated with arginine, coupled with the failure to demonstrate the metabolite in the urine of patients suspected of having arginine deficiency resulting from genetic defects of the urea cycle, suggests that arginine is an intermediate in guanidosuccinic acid synthesis. (24 refs.) - *Journal abstract.*

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- 2333 SOLITARE, G. B., SHIH, V. E., NELLIGAN, D. J., & DOLAN, T. F., JR. Argininosuccinic aciduria: Clinical, biochemical, anatomical, and neuropathological observations. *Journal of Mental Deficiency Research*, 13(3): 153-170, 1969.

Mental retardation as well as other neurological manifestations in argininosuccinic aciduria could be caused by arginine deficiency or could be the result of the toxic effect of high levels of argininosuccinic acid on the CNS. Assays of urea cycle enzymes in the liver of an 8-month-old female with this disease suggest a deficiency of liver enzymes; free amino acids in the liver are normal except for increases in glutamine, citrulline, and ornithine. Argininosuccinase was low in the liver, and argininosuccinic acid was found in the liver and brain. A defect common in amino acid metabolic disorders in which MR is a clinical occurrence is the abnormal production and/or maintenance of myelin. In this case, the basic neuropathological changes did include myelin deficiencies over large areas and a large amount of astrocytic nuclear swelling. (41 refs.) - R. L. Butler.

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- 2334 GERBER, MARCIA G., & GERBER, DONALD A. A simple screening test for histidinuria. *Pediatrics*, 43(1):40-43, 1969.

Cases of histidinemia and histidinuria can be identified easily because of the ability of excess histidine in an aliquot of urine to inhibit the formation of a blue color when cuprizone is added to a copper solution. This reaction is based on histidine itself and not

on its metabolite, imidazolepyruvic acid, which is unreliably detected by traditional ferric chloride and Phenistix tests. Histidine concentrations greater than 20 mg/100 ml can be identified and serial urine dilutions can give a semiquantitative estimate of the true concentration. No false negatives are known. This simple technique can be used to screen large numbers of children in order to identify early cases of histidinemia. (13 refs.) - E. L. Rowan.

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- 2335 MORROW, GRANT, III, BARNES, LEWIS A., AUERBACK, VICTOR H., DIGEORGE, ANGELO M., ANDO, TOSHYUKI, & NYHAN, WILLIAM L. Observations on the coexistence of methylmalonic acidemia and glycinemia. *Journal of Pediatrics*, 74(5):680-690, 1969.

Examination of 5 new published cases of methylmalonic acidemia showed that it is characterized clinically by lethargy, persistent vomiting, ketoacidosis, neutropenia, failure to grow, and MR. Methylmalonic acidemia is indistinguishable clinically from ketonic glycinemia; however, glycinemic patients do not excrete methylmalonate in their urine. Nevertheless, glycinemia was found in 8 cases of methylmalonic acidemia. Genetic studies on 10 families of children with this disorder indicate that it is apparently transmitted as an autosomal recessive trait. (23 refs.)
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- 2336 MORROW, GRANT, III, & BARNES, LEWIS A. Studies in a patient with methylmalonic acidemia. *Journal of Pediatrics*, 74(5):691-698, 1969.

A severely physically and mentally retarded 21-month-old child with methylmalonic acidemia was found to have high levels of methylmalonate in the urine (565-4850 mg/day), high plasma methylmalonate (30 mg/100 ml), and glycinemia (0.9-7.7 mg/100 ml). Glycine metabolism appeared normal based on: normal urinary levels of creatine, creatinine, and uric acid; adequate plasma serine, red cell glutathione and blood Δ -amino levulinic acid formation; and rapid excretion of administered

doses of benzoate. The only apparent abnormality of the glycine pathway was hypo-oxaluria. Restriction of protein intake decreased methylmalonate excretion, and bicarbonate administration decreased acidosis; however, no improvement occurred in his clinical condition. Administration of vitamin B₁₂, cholestyramine resin^{R1} (Cuemid), clofibrate^{R2} (Atromid), or pantothenate was also without effect. The patient also suffered a mild anemia of unknown etiology. (18 refs.) - L. S. Ho.

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2337 COWARD, R. F., & SMITH, P. Urinary excretion of 4-hydroxyphenyllactic acids and related compounds in man, including a screening test for tyrosyluria. *Biochemical Medicine*, 2(3):216-226, 1968.

Tyrosyluria associated with post-operative stress in 22 Ss undergoing major surgical operations occurred in all except one S (in which 4-hydroxyphenyllactic acid (PHPL) levels were elevated in the urine). PHPL was not detected in any of the control preoperative urines. Tyrosine, *p*-hydroxyphenylpyruvic acid and 4-hydroxy-3-methoxyphenyllactic acid were excreted in the urine of these Ss. In a case of malignant pheochromocytoma, 3 cases of melanoma and 8 cases of neuroblastoma, increased excretion of homovanillic acid occurred, and clinical evidence indicated that these Ss produced excessive 3,4-dihydroxyphenylalanine (DOPA). In 4 cases of gross tyrosyluria and one case each of phenylketonuria, rheumatoid arthritis, cancer, jaundice, coeliac disease and gross kidney deficiency, DOPA production was normal. In all Ss except the S with gross kidney deficiency, PHPL excretion was abnormally high. In ill children, a temporary tyrosyluria may coincide with symptoms resembling tyrosinosis. In normal male Ss in conditions of stress, such as serious injury or burns, high excretions of PHPL were noted; therefore, stress may account for the tyrosyluria found in ill Ss. A rapid screening test for PHPL was devised for urine from 10 normal Ss (with and without added PHPL) with chromatographic Whatman Number 1 paper using a solvent system containing isobutyl methyl ketone:dioxan:pyridine:water and a little extra pyridine if needed. (17 refs.) - F. J. McNulty.

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2338 SCHNEIDER, JERRY A., BRADLEY, KATHRYN H., & *SEEGMILLER, J. EDWIN. Transport and intracellular fate of cysteine-³⁵S in leukocytes from normal subjects and patients with cystinosis. *Pediatric Research*, 2(6): 441-450, 1968.

Leukocytes from 9 children (CA 5 mos-9 yrs) took up twice as much cysteine-³⁵S as did leukocytes from 9 control children. About 40% of ³⁵S was found as cystine-³⁵S in cystinotic leukocytes as compared to 2% in controls. The leukocytes were incubated with cysteine-³⁵S and the distribution ratio was calculated for intracellular to extracellular radioactivity. The distribution ratio for the patients was 34 compared with 21 for the controls. After 90 minutes incubation, the intracellular metabolites of cysteine-³⁵S showed that 30% of the ³⁵S was present as glutathione ³⁵S-N-ethylmaleimide (NEM) in both cystinotic and normal leukocytes, but the percentage present as cysteine-NEM in normal cells was twice that in cystinotic cells. The efflux of ³⁵S from cells after loading the cells by incubation with cysteine-³⁵S was 1% loss/minute in leukocytes from 6 patients with cystinosis compared with 0.92% loss/minute in leukocytes from 5 control Ss. The increased uptake of cysteine by cystinotic leukocytes could indicate a primary defect in the transport of cysteine in cystinosis; however, the appearance of the extra ³⁵S as intracellular cystine-³⁵S suggests that this rapid conversion may actually be the primary defect. (41 refs.) F. J. McNulty.

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2339 HOLTZAPPLE, PHILIP G., BOVEE, KENNETH, KEA, CLAIRE F., & SEGAL, STANTON. Amino acid uptake by kidney and jejunal tissue from dogs with cystine stones. *Science*, 144(3912):1525-1527, 1969.

Cystine and lysine accumulation *in vitro* in intestinal and renal tissue was studied in 8 dogs that form cystine stones. Under conditions which demonstrate *in vitro* defects in tissue obtained from humans with cystinuria, normal amino acid accumulation occurred in 6 dogs with the canine disorder. Normal amino acid uptake in tissue and the demonstration of normal to minimum increases in excretion of lysine suggest that the canine disorder is not similar to the human syndrome. (5 refs.) *Journal abstract.*

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2340 FARDEAU, MICHAEL, & *ENGEL, W. KING.

Ultrastructural study of a peripheral nerve biopsy in Refsum's disease. *Journal of Neuropathology and Experimental Neurology*, 28(2):278-294, 1969.

In a peripheral nerve biopsy examined with an electron microscope in Refsum's disease, the cytoplasm of the Schwann cells was observed to have lipoid deposits and crystalline-like bodies. There was a close relationship of the morphological lesion observed in Refsum's disease to that of Dejerine-Sottas disease. In transverse section there was a severe reduction in the number of myelinated fibers and many nerve fibers were replaced by Schwann cell proliferations formed into typical "onion-bulb" whorls. The same extensive whorling pattern is seen in Refsum's and in Dejerine-Sottas diseases. Connective tissue proliferation may be responsible for some nerve hypertrophy; however, the whorled processes are clearly from Schwann cells. The lipoid granules found in the Schwann cytoplasm are lipofuscin, lysosomes, and others that show some similarities to those of peripheral nerve metachromatic leukodystrophy granules. The crystalline-like bodies are unusual in peripheral nerve pathology. Frequently, the light microscopic examination of peripheral nerve biopsies of Refsum's disease has been disappointing; therefore, for hypertrophic neuropathies, phase contrast or paraphenylenediamine staining gives the best visibility of the delicate whorls. (40 refs.)
F. J. McNulty.

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2341 GORDON, NEIL. Metabolic disorders affecting vision. In: Gardiner, Peter, MacKeith, Ronald, & Smith, Vernon, eds. *Aspects of Developmental & Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p. 91-95.

Metabolic diseases which affect the eye include homocystinuria, Wilson's disease, galactosemia, Refsum's disease, and 6 mucopolysaccharidosis syndromes (Hurler's, Hunter's, Sanfilippo, Morquio, Scheie, and Maroteaux-Lamy). Clinical symptoms in homocystinuria (an error in the metabolism of sulfur containing amino acids) include MR, ectopia lentis, iridodonesis, and livedo reticularis among other more well-known characteristics. Demonstration of Kayser-Fleischer rings in the eyes substantiates a diagnosis of Wilson's disease, and oral penicillamine is generally

used for treatment. Almost 1/2 of the children who are untreated for galactosemia develop zonular or lamella cataracts. The main clinical features of Refsum's syndrome are, ataxia, hemeralopia, concentric limitation of the visual fields, and polyneuritis. Clouding of the cornea and the lumbar gibbus are typical of Hurler's, but not Hunter's, syndrome. Retinitis pigmentation may lead to blindness in the Sanfilippo syndrome, and corneal clouding appears both in the Morquio and Schreie syndromes. (8 refs.) - V. G. Votano.

2342 DOUGLAS, A. A. The eye in cystine storage disease with a note on the use of the slit lamp. In: Gardiner, Peter, MacKeith, Ronald, & Smith, Vernon, eds. *Aspects of Developmental & Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p. 96-100.

A slit-lamp examination is usually required to observe the primary crystalline degeneration of the cornea which occurs in intermediate and severe cases of cystine storage disease. The conjunctiva contains chalky-looking deposits which are numerous in the fornices and at the limbus. Confirmation of the disease can be made by examining lymph node biopsies for crystalline material. The disease appears to be inherited as a simple Mendelian recessive character (ratio of unaffected to affected 3:1). The medium in which the crystal is found has a profound effect on its shape. In tissues, they are acicular or prismatic, and their plane is perpendicular to the long crystal axis and to the optical axis. (5 refs.) - V. G. Votano.

2343 Glycogen storage disease. *British Medical Journal*, 2(5649):68, 1969.
(Editorial)

Clinically similar forms of the hepatomegalic type of glycogen storage disease are caused by the congenital absence of at least 3 different enzymes: glucose-6-phosphate (Type I or von Gierke's disease), amylo-1, 6-glucosidase (Type III), and phosphorylase (Type VI). Glycogen assay in liver biopsy material may be supplanted by blood cell or intestinal mucosa cell enzyme assay in the definitive diagnosis. Rational defensive dietary therapy is possible once the mechanism of the abnormality is known. (11 refs.) - E. L. Rowan.

- 2344 OCKERMAN, PER-ARNE. Diseases of glycoprotein storage. *Lancet*, 1(7597): 734, 1969. (Letter)

A patient with progressive psychomotor retardation showed a deficiency of α -mannosidase activity and intense storage of material containing mannose and glucosamine. Like aspartyl-glucosaminuria, this mannosidosis is classified as a disease of glycoprotein storage and is separated from both glycosaminoglycan and glycolipid storage diseases even though glycoprotein storage may occur in these latter syndromes. (14 refs.) - E. L. Rowan.

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- 2345 KALCKAR, HERMAN M. Development of anabolic pathways of galactose and glucose metabolism in man. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thoms, 1968, Chapter 5, p. 35-39.

The pathophysiological development of the lens, liver, and brain in hereditary galactosemia is probably caused by the accumulation of galactose 1-phosphate. Diagnosis of hereditary galactosemia requires careful enzymatic analysis which begins with a demonstration of galactose 1-phosphate accumulation or lack of galactose metabolism and then investigates possible defects of other enzymes in the pathway. Since the enzymatic defect is limited to the enzyme galactose 1-phosphate uridyl transferase, the institution of a galactose-free diet as soon as possible after birth prevents cellular damage. Studies of the enzyme, uridine diphosphate (UDP)-glucose dehydrogenase, and the enzyme which catalyzes the transfer of UDP-glucuronic acid to bilirubin indicate that the sluggish rise of these enzymes may be responsible for the problems which arise in the case of blood-group incompatibility. When an increasing amount of free bile pigments are liberated and deposited in lipophilic areas as in erythroblastosis fetalis, it may be desirable to start the detoxification mechanism early to prevent kernicterus and SMR. Research is needed to uncover ways to start a detoxification process which would elicit high levels of UDP-glucose dehydrogenase, UDP-glucuronic acid, and the succeeding enzymes immediately after birth. (24 refs.) - J. K. Wyatt.

- 2346 DAHLQVIST, A., JAGENBURG, R., & MARK, A. A patient with hereditary galactosemia studied with a screening method for galactose in urine. *Acta Paediatrica Scandinavica*, 58(3):237-244, 1969.

Hereditary galactosemia in a newborn infant was diagnosed on the basis of the presence of galactose in the blood and urine, and the absence of galactose-1-phosphate uridyl transferase activity in red blood cells. This enzyme activity was about 50% of the normal value in both parents, suggesting that they are heterozygous for hereditary galactosemia. A galactose-free diet was introduced on the ninth day of life, and the patient rapidly improved. Only a slight retardation which affects all functions equally remains as sequelae of the initial galactosemia. A new simple test paper for galactose was used to estimate galactose content in the urine and to check the suitability of the diet. In addition to galactose, galactitol was found in the urine. Urine galactose and galactitol, aminoaciduria, and increased blood amino acid concentrations disappeared on the galactose-free diet. (44 refs.) - L. S. Ho.

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- 2347 HOLZEL, ARON. Galactosemia. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 13, p. 823-829.

Galactosemic newborns on a milk diet (containing lactose) develop vomiting, refusal to feed, listlessness, jaundice, diarrhea, and loss of weight in the first week of life. If dietary treatment is delayed, liver enlargement, hemorrhage, and hyperproteinemia may follow, and liver dysfunction or intercurrent infections may lead to death. Cataracts and brain damage can appear in surviving patients; however, early diagnosis and rigid exclusion of galactose from the infant's diet should ensure normal development, provided intrauterine brain injury has not occurred. The relatively high incidence in male offspring, and the absence of the disease in parents suggest that galactosemia may be transmitted by a single recessive autosomal gene. Determination of galactose-1-phosphate uridyl transferase in red cells is a reliable assay for galactosemia; normal cells produce 5.9 uridine diphosphoglucose/ml erythrocytes/hour; in galactosemic cells the value is 0.1; and in heterozygote carriers the value is 3.0. (30 refs.) - L. S. Ho.

2348 SIDBURY, JAMES B., JR. The glycogenoses. In: Gardner, Lytt, I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 13, p. 853-867.

Hereditary glycogenoses can be classified into 9 groups on the basis of the enzyme defects involved in the abnormal synthesis and storage of glycogen: glucose-6-phosphatase (type I), acid α -1,4-glucosidase (type II), amylo-1,6-glucosidase and/or oligo-1,4-1,4-glucantransferase (type III), amylo-1,4-1,6-transglucosylase (type IV), muscle phosphorylase (type V), liver phosphorylase (type VI), phosphofructokinase (type VII), phosphohexoisomerase (type VIII), and phosphorylase kinase (type IX). Type I, II, and VI are clinically similar; the affected infants may have mild hepatomegaly, respiratory distress, hypoglycemic convulsions, and ketonuria. Type IV exhibits clinically distinct hepatic cirrhosis. Type II, V, and VII primarily involve the muscle and type II affects infants primarily. Treatment includes high carbohydrate feedings between meals for hypoglycemia for type I, a high protein diet for types III and VI. No effective treatments have been found for types II and V. Very few cases are reported of types IV, VII, VIII, and IX. Aglycogenesis due to glycogen synthetase deficiency shows clinical pictures similar to types I and III. Intravenous administration of glucose and high carbohydrate diets offer good protection against hypoglycemic seizures. (68 refs.) - L. S. Ho.

2349 CARAKUSHANSKY, GERSON. Disorders of heme and porphyrin metabolism. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 20, p. 994-1003.

The more serious forms of familial and congenital nonhemolytic jaundice are: Crigler-Najjar syndrome (associated with early onset, severe neurological signs, and short life-span); fetal familial intrahepatic cholestasis (Byler's disease) (associated with hepatosplenomegaly, dwarfism, and early death); and transient familial hyperbilirubinemia. Treatments of choice are exchange transfusions and cholestyramine administration. Gilbert's syndrome (constitutional hepatic dysfunction) and Dubin-Johnson syndrome (chronic idiopathic jaundice with abnormal pigment in hepatic cells) are the milder forms of jaundice. No treatment is required and prognosis is good in these disorders. Acute intermittent porphyria is the most severe form of familial porphyrin metabolic disorders and is often associated with muscular paralysis and

neurotic and psychotic manifestations. Congenital erythropoietic porphyria, and porphyria cutanea tarda hereditaria are characterized by the skin sensitivity to sunlight. (57 refs.) - L. S. Ho.

2350 LOEB, H., TONDEUR, M., TOPPET, M., & CREMER, N. Clinical, biochemical and ultrastructural studies of an atypical form of mucopolysaccharidosis. *Acta Paediatrica Scandinavica*, 58(3):220-228, 1969.

The clinical features (MR, inconspicuous facial and bone deformities, absence of corneal clouding, hepatosplenomegaly, stiffness of the joints, and presence of cytoplasmic vacuoles in lymphocytes) of a boy and his sister fit well with that of mucopolysaccharidosis type III; however, the radiological and biochemical data do not agree with the clinical findings. Typical bone lesions are not present in the girl. Both heparitin sulfate and chondroitin sulfate B were found in the urine; however, the amount of the urinary mucopolysaccharides expressed in milligrams of uronic acid was normal in the girl. Some liver acid hydrolases were higher than normal, a frequent observation in gargoylism. In contrast to usual cases, a strikingly high acid β -galactosidase activity level was observed. Liver ultrastructure showed swollen lysosomes with complex storage of lipids and mucopolysaccharides. (41 refs.) - L. S. Ho.

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2351 EHRLICH, ROBERT M., & MARTIN, JULIO M. Diazoxide in the management of hypoglycemia in infancy and childhood. *American Journal of Diseases of Children*, 177(4): 411-416, 1969.

Oral administration of diazoxide was effective in raising blood sugar levels in 5 of 9 children with idiopathic hypoglycemia. Four of the 9 hypoglycemic children responded to diazoxide with a growth spurt, improved glucose tolerance, and decreased sensitivity to oral leucine. An increase in body hair was observed in 3 cases, but this disappeared when the drug was stopped. One child developed vomiting, another had facial edema, and neither responded to diazoxide treatment. One of the 9 patients died with hyperglycemia and bronchopneumonia. A safe range of drug dosage appears to be 10-15 mg/kg/day orally with

careful monitoring of all patients.
(14 refs.) - L. S. Ho.

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2352 HENDERSON, J. FRANK, KELLEY, WILLIAM N., ROSENBLOOM, FREDERICK M., & *SEEG-MILLER, J. EDWIN. Inheritance of purine phosphoribosyltransferases in man. *American Journal of Human Genetics*, 21(1):61-70, 1969.

A complete loss of hypoxanthine-guanine phosphoribosyltransferase is associated with a syndrome characterized by self-mutilation, MR, spasticity, and choreoathetosis, while a partial loss is found in patients with gouty arthritis. A partial loss of adenine phosphoribosyltransferase was found in 4 healthy members in 3 generations of one family. Pedigree analyses of 2 families with complete or partial loss of hypoxanthine-guanine phosphoribosyltransferase indicated an X-linked recessive mode of inheritance with a close linkage between X_g and the locus for hypoxanthine-guanine phosphoribosyltransferase. A comparison of thermostability of adenine phosphoribosyltransferase in several persons with a partial loss of the enzyme and in normal controls suggests the occurrence of at least 2 types of enzyme differing in thermostability (due perhaps to alterations in the structural gene). There was no evidence of sex limitation or sex linkage, and the alleles involved are probably autosomal, thus involving a chromosome different from the one containing the gene for hypoxanthine-guanine phosphoribosyltransferase. (13 refs.) - M. G. Conant.

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2353 HAUST, M. DARIA, ORIZAGA, MANUEL, BRYANS, ALEX M., & FRANK, HAROLD F. The fine structure of liver in children with Hurler's syndrome. *Experimental and Molecular Pathology*, 10(2):141-161, 1969.

Although Hurler's syndrome is classically associated with storage of mucopolysaccharide,

recent electron microscopic studies of the liver may require revision of this concept. Hepatic tissue was obtained from 7 children with clinical evidence of Hurler's syndrome and from 2 controls. The most characteristic findings in Hurler's syndrome were sinusoidal dilations and vacuoles in the parenchymal cytoplasm. Vacuoles arise, at least in part, from the breakdown of organelles (mitochondrial budding) to form vesicles which grow and disrupt cellular architecture. As disease progresses, free-floating myelin bodies and "zebra bodies" appear in the sinusoids. These have previously been reported only in nervous tissue and are composed of gangliosides. The basic error must be in enzyme systems involved in the metabolism of both gangliosides and mucopolysaccharides. (60 refs.) - E. L. Rawan.

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2354 MacBRINN, MONICA, OKADA, SHINTARO, WOOLLACOTT, MARJORIE, PATEL, VIMAL, HO, MAE WAN, TAPPEL, A. L., & O'BRIEN, J. S. Beta-galactosidase deficiency in the Hurler syndrome. *New England Journal of Medicine*, 281 (7):338-343, 1969.

A deficiency of β -galactosidase (pH 5.0) was found in frozen tissues (brain, liver, kidney and spleen) from 10 patients with Hurler's syndrome (Types 1-3). The diminished activity of this enzyme was demonstrated with the use of nitrophenylgalactosides as well as ganglioside GM₁ and a "keratan sulfate-like" mucopolysaccharide. Mixing experiments demonstrated that soluble endogenous inhibitors are not responsible for the lowered β -galactosidase activity. Activities of related hydrolases, including acid phosphatase, α -glucosidase, β -galactosidase, β -xylosidase, β -N-acetyl-galactosaminidase, β -N-acetylglucosaminidase, α -mannosidase, β -glucuronidase and cathepsins A-D, were increased 2 to 5 times. Deficiency of β -galactosidase may prove to be the primary enzymic defect in the Hurler syndrome. (27 refs.) - *Journal abstract.*

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2355 MATALON, REUBEN, DORFMAN, ALBERT, DAWSON, GLYN, & SWELEY, CHARLES C. Glycolipid and mucopolysaccharide abnormality in fibroblasts of Fabry's disease. *Science*, 164 (3887):1522-1523, 1969.

Analysis of skin fibroblast cultures from a man with Fabry's disease revealed an accumulation of the glycolipid galactosyl-galactosyl-glucosyl ceramide (GL-3) which had previously been found in other tissues. There was no abnormal mucopolysaccharide as is commonly found in storage disease, but there was an accumulation of normal acid mucopolysaccharides and marked metachromasia was noted when the cultures were stained with toluidine blue. This evidence suggests that there is an absence of the enzyme that hydrolyzes the terminal D-galactose residue of the deposited trihexosylceramide and that this enzyme is specific for glycolipids and not for mucopolysaccharides. (18 refs.) - E. L. Rowan.

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2356 JOSEPH, M. C., & *MEADOW, S. R. The metacarpal index of infants. *Archives of Disease of Childhood*, 44(236):515-516, 1969.

The metacarpal index, the sum of the lengths of second, third, fourth, and fifth metacarpals divided by the sum of the breadths of the same fingers, was greater in children with Marfan's syndrome or arachnodactyly than in normal children of comparable age. The standards were obtained in 50 normal children at the ages of 6, 12, 18, and 24 months. The index increases with age and is greater in girls than in boys. High indices correlate with long fingers. Mongoloids who usually have short fingers, nevertheless, have normal indices. The metacarpal index may be helpful in diagnosing Marfan's syndrome, often difficult in early life. (4 refs.)
L. S. Ho.

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2357 BOONE, JOHN A., & CLOWES, GEORGE H. A., JR. Calcified annulus fibrosus with mitral insufficiency in the Marfan syndrome. *Southern Medical Journal*, 62(6):682, 690, 1969.

A 24-year-old male with a high, narrow palatal arch and long hands and feet suggestive

of the Marfan syndrome had a grade 3 to 4 harsh systolic murmur, mitral insufficiency, and heavy calcification of the mitral annulus. A Starr-Edwards ball valve prosthesis was inserted, and postoperative improvement was steady following electrical reversion of atrial fibrillation. Three and one-half years after the operation, a grade 1/6 apical systolic murmur remained, but the patient was working full time and remaining asymptomatic. (5 refs.) - M. G. Conant.

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2358 ROSENBERG, LEON E. Inherited amino acidopathies demonstrating vitamin dependency. *New England Journal of Medicine*, 281(3):145-153, 1969.

Inherited vitamin-dependency states are defined as "genetic disturbances that lead to specific biochemical abnormalities affecting only one reaction catalyzed by a vitamin and that respond only to pharmacological amounts of the vitamin." Both vitamins B₆ and B₁₂ have been implicated in specific inherited defects of amino acid metabolism which are responsive to vitamin administration; both have important roles in amino acid metabolism. Five disorders with demonstrable vitamin B₆ dependency are known: infantile convulsions; pyridoxine-responsive anemia; cystathioninuria; xanthurenic aciduria; and homocystinuria. Each condition has distinct clinical and biochemical findings, each is familial, and each results from a specific error of metabolism. Administration of massive doses of vitamin B₁₂ to a child with methylmalonic aciduria and with no other signs of vitamin B₁₂ deficiency resulted in a fall in methylmalonic acid excretion. This suggests that the illness is a dependency state rather than a deficiency state. (40 refs.) - M. G. Conant.

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2359 TURNER, DAVID ALAN, & CEVALLOS, WILLIAM H. Vitamin B₁₂ and the lipids of the central nervous system. *Clinical Biochemistry*, 2(1):1-11, 1968.

A successive generation of rats maintained on a vitamin B₁₂ deficient diet did not develop neurological symptoms similar to those of man after prolonged vitamin B₁₂ deprivation. The rats did not follow a normal growth curve of weight gain and had lower levels of vitamin

B₁₂ in the plasma, liver, kidney, and spleen compared with the controls; however, histological examination of nerve and brain failed to reveal any changes in the myelin of these tissues. The lipid concentration of the sciatic nerve showed a decrease in the triglyceride concentration. The phospholipid levels did not change significantly; however, the rate of turnover may have been reduced. The total incorporation of labeled phosphorus into the phospholipid fraction of the rat brain, spinal cord, and sciatic nerve was discovered to be a slow process. (22 refs.)

F. J. McNulty.

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2360 McCANDLESS, DAVID W., & *SCHENKER, STEVEN. Encephalopathy of thiamine deficiency: Studies of intracerebral mechanisms. *Journal of Clinical Investigation*, 47(10):2268-2280, 1968.

The pyruvate decarboxylase and transketolase activity, lactate, adenosine triphosphate (ATP), and reduced glutathione (GSH) levels in the cerebral cortex, cerebellum, and brainstem and thiamine concentration in whole brain of rats with diet-induced thiamine encephalopathy were measured and compared with those of pair-fed and normally fed controls. Definite neurological signs appeared in the experimental animals only after 4.5-5 weeks of thiamine deprivation when the brain thiamine level had fallen to less than 20% of normal. Administration of thiamine hydrochloride resulted in immediate (16-36 hours) reversal of neurological signs when the brain thiamine level had reached only 26% of normal. Apparently, cerebral transketolase and pyruvate decarboxylase activities are impaired, primarily in the brainstem and the cerebellum, but both rise on reversal of the neurological state. Cerebral ATP concentration is normal, and GSH concentration is slightly decreased during encephalopathy. Transketolase depression probably reflects another critical aspect of the hexose monophosphate shunt. (42 refs.) - M. G. Conant.

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2361 KOCHHAR, D. M. Studies of vitamin A-induced teratogenesis: Effects on embryonic mesenchyme and epithelium, and on incorporation of H³-thymidine. *Teratology*, 1(3):299-310, 1968.

Oral vitamin A treatment of pregnant rats on days 10, 11, and 12 of gestation (60,000 IU/day) markedly retarded the growth of the embryos and inhibited ³H-thymidine incorporation by the liver, the cephalic mesenchyme tissue, the posterior region of the palatine shelves, trigeminal nucleus, and some skin tissues. Initial defects of embryos were found in certain facial tissues, while the overall organogenesis progressed normally. Mesodermal cells in the maxillary process produced cartilaginous elements, while in control embryos the maxillary bone was developed from the same tissue later on and was without an intermediate cartilage. Oral epithelium, especially near the sites where chondrogenic tissue is induced in the mesoderm, showed proliferations usually associated with the development of glands. The palatal and dental abnormalities in vitamin A-treated embryos are probably related to the changes in the epithelium and the altered epithelial-mesodermal interaction. (24 refs.) L. S. Ho.

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2362 GARDNER, LYTT, I. Historical notes on cretinism. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 3, p. 212-215.

The earliest important scientific observation on cretinism was described by Paracelsus (1603) in connection with endemic goiter. The first 2 cases of sporadic, athyroidic cretinism were published by Thomas Curling (1850). Gordon (1922) observed 280 cases of sporadic cretinism (called childhood myxedema) in North America and noted that twice as many females as males were affected and that hereditary influences had little importance. The most recent definition of cretinism found in Webster's Collegiate Dictionary (1967) is "a usually congenital abnormal condition marked by physical and mental stunting and caused by severe thyroid deficiency." Child and Gardner (1954) noted that birth-weights of cretins were greater than normal and that frequency distribution for the cretins showed a bimodal curve, suggesting there are 2 groups of cretins, one with normal birth-weights and the other with greater than normal birth-weights. (18 refs.) - L. S. Ho.

- 2363 ANDERSON, HENNING J. Nongoitrous hypothyroidism. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 3, p. 216-234.

The somatic growth and development of children with congenital nongoitrous hypothyroidism can usually be corrected by administration of thyroid hormones; however, the mental outcome is dependent upon the age treatment is instituted. Best results may be obtained by giving continuous full therapy as early as possible. There are very few signs to alert physicians to athyrosis in fetal or in early postnatal life, the most important period of cerebral maturation. Hypothyroidism after the third year of life does not cause irreparable damage to the brain. The etiology, clinical symptoms, differential diagnosis, and treatment of congenital nongoitrous hypothyroidism need further elucidation. (106 refs.) - L. S. Ho.

- 2364 KOENIG, M. PIERRE. Endemic goiter and endemic cretinism. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 3, p. 235-243.

Endemic goiter is widely distributed over the world, and the occurrence of endemic cretinism coincides with severe endemic goiter centers. The incidence of endemic cretinism can be as high as 8% of the population in goiter regions such as Mulia, New Guinea, and certain parts of equatorial Andes. Iodine supplementation has proven to be remarkably effective in preventing and alleviating goiter. However, factors other than iodine deficiency such as hereditary characteristics and goitrogenic agents play a part in the genesis of endemic goiter. The clinical picture of endemic goiter is almost indistinguishable from that of sporadic nontoxic goiter. The most prominent symptoms of endemic cretinism are mental deficiency, auditory and neurologic defects, and skeletal stunting. Endemic cretinism may or may not be accompanied by hypothyroidism. Endemic cretins should be treated with thyroid hormones if there are hypothyroid symptoms; however, neurological disorders are usually irreversible. (33 refs.) - L. S. Ho.

- 2365 HUTCHISON, JAMES H. Familial goitrous hypothyroidism. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 3, p. 253-267.

Hereditary goitrous hypothyroidism usually responds to continuous administration of thyroxine with great physical and clinical improvement; however, intellectual improvement is less satisfactory unless the hypothyroid state is diagnosed in early months of life. Hypothyroidism before the second year of life has been shown to damage brain development irreversibly; therefore, early diagnosis is extremely important. Thyroidectomy should be performed if the gland contains palpable nodules in order to prevent malignant change in the hyperplastic tissues. Familial goitrous hypothyroidism can be classified into 5 types: an iodine transport-mechanism defect, an iodotyrosine coupling-mechanism defect, a thyroid peroxidase deficiency, a thyroid dehalogenase deficiency, and an abnormal iodinated protein production. Deficiency of peroxidase can be established by the radioactive iodine technique. Differential diagnosis of other types of intrathyroid metabolic defects is not possible at the present time. Alternative treatments other than administration of thyroxine are administration of potassium iodide and triiodothyroacetic acid. (76 refs.) L. S. Ho.

- 2366 SCHULMAN, JOSEPH D., & *CRAWFORD, JOHN D. Congenital nystagmus and hypothyroidism. *New England Journal of Medicine*, 280(13):708-710, 1969.

Congenital nystagmus and hypothyroidism were found in 4 patients (1 MR); this association has not been noted previously. Since separate occurrences of congenital nystagmus and hypothyroidism are not frequent, it is not likely that the association noted here is due to chance alone. This association could hasten the diagnosis and early treatment of the metabolic disorder. (18 refs.) - L. S. Ho.

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- 2367 KLEIN, ROBERT. Hypoparathyroidism. In: Gardner, Lytt, I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 4, p. 373-391.

Idiopathic hypoparathyroidism occurs more frequently in children than adults and presents symptoms of hypocalcemia, convulsions, and tetany. Mental status in such children is difficult to establish because of repeated convulsions; however, intellectual ability usually improves to normal with treatment. Metastatic calcification and change of ectodermal structure are probably the major symptoms directly resulting from the lack of parathyroid hormone. Many cases of neonatal and infantile hypoparathyroidism seem to be inherited by a sex-linked recessive trait; all these patients are boys. Other syndromes of hypoparathyroidism are transient congenital hypoparathyroidism, congenital absence of the parathyroid glands, and idiopathic hypoparathyroidism of later onset. Pseudohypoparathyroidism which occurs twice as frequently in females as in males can be diagnosed by its non-response to parathyroid hormone. The presence of brachydactylia, short stature, moon face, subcutaneous calcification, MR, and family history of the syndrome is also useful in differential diagnosis. Maintaining a normal serum calcium level is most important in both conditions. Parathyroid hormone is not used because it has to be administered parenterally and because it may cause hypersensitivity. (88 refs.) - L. S. Ho.

results from a chronic lack of stimulation for its release. Total thyroidectomy is not warranted in pseudohypoparathyroidism. (31 refs.) - L. S. Ho.

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- 2369 HERTZOG, KEITH P. Brachydactyly and pseudo-pseudohypoparathyroidism. *Acta Geneticae Medicae et Gemellologiae*, 27(3): 428-437, 1968.

Brachydactyly of the fifth middle phalanges was found in 12 of 96 Chinese children (CA 4-16 yrs) in Philadelphia. In 9 of these Ss, stunted cratered diaphyses with precocious cone epiphyses were found. Epiphyseal union also occurred precociously in these children. Cases of metacarpal brachydactyly can be divided into 2 distinct subgroups: one in which the brachydactyly is limited to the fourth metacarpal and/or metatarsal and one which is characterized by a variable combination of metacarpals. Phalangeal involvement is rare in pseudo-pseudohypoparathyroidism; therefore, these cases probably belong in another classification. (47 refs.) - M. G. Conant.

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- 2368 SUH, SE MO, KOOH, SANG WHAY, CHAN, ALICE M., *FRASER, DONALD, & TASHJIAN, ARMEN H., JR. Pseudohypoparathyroidism: No improvement following total thyroidectomy. *Journal of Clinical Endocrinology and Metabolism*, 29(4):429-439, 1969.

Total thyroidectomy in an 11-year-old MR girl with pseudohypoparathyroidism did not correct the hypocalcemia and refractoriness to exogenous bovine parathyroid hormone. The thyroid gland was removed in 2 stages while serum calcium was maintained by intravenous infusion of calcium. The calcitonin content of the right lobe of the thyroid was 0.60 MRC unit/gm (10-20 times normal) and that of the left lobe was 0.35 MRC unit/gm. Since the patient remained refractory to bovine parathyroid extract, the high calcitonin content of the thyroid is probably not the cause of hypocalcemia, but a secondary effect which

- 2370 LATHAM, S. C., BORDIER, PH., DOYLE, F. H., HIGGS, F. D., JOPLIN, G. F., TAYLOR, SELWYN, & THALASSINOS, N. C. A case of mild hyperparathyroidism in childhood. *Archives of Disease in Childhood*, 44(236):521-526, 1969.

Transient hyperparathyroidism in a 9-year-old girl was discovered to be due to a small parathyroid adenoma. Removal of the tumor restored the phosphate excretion index, urinary calcium, and plasma calcium and phosphate to normal. The child was originally admitted to the hospital with abdominal pain, vomiting, intense constipation, and headaches. Laboratory tests revealed hypercalciuria and moderate hypercalcemia which could be suppressed by oral hydrocortisone. Later, oral hydrocortisone was only partially effective. Her condition was determined not to be from vitamin D hypersensitivity or intoxication, and sarcoidosis was also excluded. The

differential diagnosis of hyperparathyroidism was made on the basis of the phosphate excretion index, bone biopsy, and measurements of radiological bone density. (18 refs.)
L. S. Ho.

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- 2371 YOUNOSZAI, M. K., KACIC, ANNE, DILLING, LOUISE, & *HAWORTH, J. C. Urinary hydroxyproline: Creatinine ratio in normal term, pre-term, and growth-retarded infants. *Archives of Disease in Childhood*, 44(236):517-520, 1969.

The urinary peptide-bound hydroxyproline/creatinine ratio is a valid index of skeletal growth during the perinatal period. The ratio in pre-term infants is significantly higher ($p < .05$) and that in growth-retarded infants is significantly lower ($p < .05$) than in normal term infants on the first day of life. By the seventh day, the normal infants showed a significantly higher ratio than the other 2 groups. Pre-term infants excreted more free hydroxyproline on the percentage basis than the normal and growth-retarded infants; therefore, total hydroxyproline is not a reliable index for skeletal growth in newborns. A 24-hour urine collection is not necessary, since the peptide hydroxyproline/creatinine ratio is the same for randomly collected urine as for 24-hour urine samples. (19 refs.) - L. S. Ho.

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- 2372 HILLMAN, DONALD A., & COLLE, ELEANOR. Plasma growth hormone and insulin responses in short children. *American Journal of Diseases of Children*, 117(6):636-644, 1969.

Plasma growth hormone (PGH) response to insulin-induced hypoglycemia, tyressin infusion, and arginine infusion was tested in 112 children (including 9 with Turner's syndrome, 4 cretins, and 1 mongoloid) with growth failure and insulin was found to be the most effective stimulant for PGH release. Sixteen children showed significantly less response to insulin than the control group; of these, 6 were hypothyroid and 10 were hypopituitary. Children with fetal malnutrition and severe renal disease had a greater than normal response to hypoglycemia. Plasma

insulin response to arginine infusion in fetally malnourished children was significantly lower than that in children with constitutional short stature or those with gonadal dysgenesis. (25 refs.) - L. S. Ho.

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- 2373 OPITZ, JOHN M., ZELLWEGER, HANS, SHAN-NON, W. R., & PTACEK, LOUIS J. The RSH syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 43-52.

Eleven additional cases of the "RSH syndrome" show many of the features first described by Smith, Lemli, and Opitz in 1964. Growth failure, "feeding difficulties," microcephaly, and profound MR are noted early in infancy. The characteristic facies show ptosis, strabismus, anteversion of the nostrils, minor ear anomalies, micrognathia, broad alveolar ridges, and whitish hair. Cataracts, pyloric stenosis, cleft palate, and a high total ridge count sometimes occur. Hypospadias is a frequent anomaly in males. Seven families have had more than one affected child, and an autosomal recessive mode of inheritance is suggested. (8 refs.) - E. L. Rowan.

- 2374 DALLAIRE, LOUIS, & FRASER, F. CLARKE. The Smith-Lemli-Opitz syndrome of retardation, urogenital and skeletal anomalies. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 180-182.

The psychomotor retardation, urogenital, and skeletal anomalies characteristic of the Smith-Lemli-Opitz syndrome were seen in 9 children in 6 sibships. Low birth-weight, odd facies, failure to thrive, short stature, MR, microcephaly, heart defects, simian creases, and syndactyly occurred quite commonly, while cleft palate, eye anomalies, pyloric stenosis, and polydactyly were less frequent. Cytogenetic and biochemical investigations revealed no specific abnormalities, but an autosomal recessive mode of inheritance seems likely. (No refs.) - E. L. Rowan.

- 2375 HALL, JUDITH G., & JASION, ARTHUR R. Thoracic asphyxiating dystrophy, the Mohr syndrome (OFD II) or a "New Entity"? In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 207-210.

A boy with dwarfism (distal shortening of limbs), a small chest, cardiac murmur, cleft lip, pedal polydactyly, hearing loss, and probable MR had recurrent episodes of aspiration pneumonitis compatible with thoracic asphyxiating dystrophy. However, the whole constellation of anomalies does not fit this entity, the Mohr syndrome, or the Ellis-van Creveld syndrome and probably represents a new dysmorphogenetic syndrome. (3 refs.)
E. L. Rowan.

- 2376 SCOTT, CHARLES I. Low birth weight dwarfism in two brothers. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 241-244.

Brothers (CA 6 1/2 and 4 1/2 yrs) showed a previously unreported identical syndrome of short stature with disproportionately short hands and feet, unusual facies (prominent, long nose, high nasal bridge, small receding mandible), small teeth but advanced dental age, and mild MR. X-ray examinations showed dislocation of radial heads, dolichocephaly, and small but proportioned bones. (No refs.)
E. L. Rowan.

- 2377 DRILLIEN, C. M. The small-for-date infants: Etiology and prognosis. *Pediatric Clinics of North America*, 17(1):9-24, 1970.

Intrauterine growth retardation is related to maternal undernutrition, maternal pathological conditions, and genetic influences. The mental and physical growth and development during childhood of these infants are poorer than well grown children, but are better than premature infants. Brain growth, function, and structure are not noticeably affected by *in utero* growth retardation. This is in contrast to the experimental results in many animal species. Although the association of poverty with a reduced birth-weight is well documented, an increase in birth-weight with improving social conditions is not noticeable. Postnatal malnutrition and environment, especially during the first year of life, affect

the physical and mental status in later years. The incidence of congenital abnormalities is increased in small-for-date infants, and those with anomalies are more likely to develop mental and neurological defects than those without anomalies. Maternal estriol excretion seems to be a good indicator of fetal-placental status. In 17 cases of below normal estriol excretion, 10 of the infants at 1 year had suspected mental or neurological defects. In 11 cases of normal estriol excretion, only one infant had a degree of MR. (42 refs.) - L. S. Ho.

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- 2378 ANDREWS, BILLY F. Amniotic fluid studies to determine maturity. *Pediatric Clinics of North America*, 17(1):49-67, 1970.

Amniocentesis with studies of amniotic fluid properties (osmolality, bilirubin, creatinine, proteins, enzymes, steroids, and cytology) seems to be the best approach for evaluation of fetal disease, gestational age, and chromosomal and metabolic disorders. Only 1-2% of the amniotic fluid is solid solutes; the solute content is comparable to that of maternal serum in early pregnancy and decreases as gestation advances. Very high osmolality is often associated with fetal diseases, distress, and abnormalities. Infants with Rh-immunization, anencephaly, intestinal obstruction, deteriorating hydrops, and diabetes show increased amniotic bilirubin levels. High amniotic protein values are found in fetal death, anencephaly, and other anomalies; however, the protein values vary widely for a given gestational age among individuals and thus cannot serve as a sole indicator for maturity. Elevation of globulins is noted in Rh-sensitized pregnancies. Creatinine and Nile blue sulfate stain of amniotic cells are fairly good indices for fetal maturity. Creatinine at 2 mg/100 ml or more and a cytological count of 10% fat-laden cells indicate maturity at 36 weeks gestation with a 95% confidence limit. No single amniotic component alone is sufficient to assess fetal maturity; nevertheless, combinations of several components, menstrual and obstetrical histories, and physical examinations should allow estimates of fetal age which have considerable accuracy. (99 refs.)
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2379 WINICK, MYRON. Cellular growth in intrauterine malnutrition. *Pediatric Clinics of North America*, 17(1):69-78, 1970.

Severe fetal malnutrition in the human, as in the rat, decreases the rate of cell division in the cerebellum, cerebrum, and brainstem, prenatally and postnatally. Human cerebrum has a higher rate of cell division postnatally than that of the rat and, therefore, is much more affected. Placental growth proceeds in parallel with organ growth in rats. Thus, changes in human placenta should give some clues to fetal growth. Placentas from infants with retarded intrauterine growth show fewer cells and a higher ribonucleic acid/deoxyribonucleic acid ratio than normal. Full-term infants who died of malnutrition during the first year showed a 15-20% reduction of total brain cells, and infants weighing 2,000 gm or less at birth who died of malnutrition had a 60% reduction. (22 refs.) - L. S. Ho.

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2380 CASSADY, GEORGE. Body composition in intrauterine growth retardation. *Pediatric Clinics of North America*, 17(1):79-99, 1970.

Plasma volume as measured by the 10-minute T-1824 albumin space is expanded in undergrown neonates. Also found are reduced cell numbers, an increased metabolic rate, and an expanded total extracellular space. Rapid adjustment to a normal plasma volume occurred by 12 hours after birth. These characteristics of body composition in cases of fetal growth retardation are similar to those of postnatal protein-calorie malnutrition. (121 refs.) - L. S. Ho.

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2381 REISMAN, LEONARD E. Chromosome abnormalities and intrauterine growth retardation. *Pediatric Clinics of North America*, 17(1):101-110, 1970.

Intrauterine growth retardation is associated with almost all syndromes with autosomal abnormalities (especially 18- and D1-trisomy) and with XO and polyploid sex chromosome disorders. Prenatal growth retardation associated with chromosome anomalies is obviously on a continuum of intrinsic genetic failure.

About 90% of abnormal conceptuses are eliminated by spontaneous abortion. Those who survive eventually fail to thrive into adulthood, and many never live normally. (34 refs.) L. S. Ho.

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2382 HUGHES, WALTER T. Infections and intrauterine growth retardation. *Pediatric Clinics of North America*, 17(1):119-124, 1970.

Low birth-weight of infants is associated with rubella, cytomegalovirus, *Herpes simplex*, toxoplasmosis, malaria, and congenital syphilis, as well as some acute bacterial infections. Immunoglobulin levels of neonates correlate well with gestational age, but not with birth-weight. Infants of low gestational age generally have a low IgG level which is often associated with pronounced hypogammaglobulinemia for the first month of life. The IgM level is normal, but the increase of IgM in the first few weeks of life is much less rapid than normal. No differences are noted in IgA and IgE. Rubella is the most well-known infection to cause intrauterine growth retardation. The virus may act directly by inhibiting fetal cell multiplication or indirectly by causing chronic ischemia due to vascular lesions in the placenta. (18 refs.) L. S. Ho.

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2383 USHER, ROBERT H. Clinical and therapeutic aspects of fetal malnutrition. *Pediatric Clinics of North America*, 17(1):169-183, 1970.

The risk of perinatal death, asphyxia, anomalies, or neonatal hypoglycemia is greatly increased in infants with birth-weights below the third percentile for their gestational age. Characteristic features of such infants are: creases on the soles of the feet; small breast nodules; coarse, straight, and silky hair; loss of fat and subcutaneous tissue; polycythemia; and acidosis and hypoglycemia which develop immediately after birth. These infants usually suck well and have good head control, unlike prematures. About 99% of the small-for-date infants show no apparent cause for the growth retardation. Possible factors in the etiology include multiple-birth pregnancy, single umbilical artery, or maternal

smoking. In addition, women who have had a previous history of giving birth to small-for-date infants or who are very thin are high risk Ss. Toxemia or infarction of the placenta are probably not important etiological factors. Most infants with fetal malnutrition are not acutely ill; however, dangerous hypoglycemia can develop. Intravenous glucose infusion immediately after birth is recommended for such infants, and the infusion should continue until milk feeding is well tolerated. Fetal malnutrition contributes to about 1.4 perinatal deaths/1,000 births. (No refs.) - L. S. Ho.

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- 2384 WALKER, JAMES. The "small for dates" baby. *Proceedings of the Royal Society of Medicine*, 60(9):877-879, 1967.

Between 24% and 35% of all births that meet the weight criteria for prematurity (less than 2,500 gm) do not meet the time criteria (are born after 37 wks gestation), and the clinical significance of these "small for dates" (SFD) babies is uncertain. There is a significant perinatal death rate for these SFD babies (10%), but the subsequent death rate in the first week is low. In 60% of the cases, the pregnancy was normal but hypertension (15%), threatened abortion (3%), and antipartum hemorrhage (5%) were sometimes present. The chances of a mother having a second SFD baby are approximately 35%. Thus, in pregnancies with a history of a previous SFD baby, hypertension, or threatened abortion, there should be a suspicion of retarded fetal growth. In many cases of SFD babies, maternal weight gain slows or stops after 30 weeks. The estriol excretion in these pregnancies continues to rise throughout pregnancy, but follows a small baby line. The degree of hazard to these SFD babies is uncertain, and classification awaits further study. (14 refs.) - W. Klein.

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- 2385 NELIGAN, G. A. The "small for dates" baby: The clinical effects of being "light for dates." *Proceedings of the Royal Society of Medicine*, 60(9):881-883, 1967.

The prognosis for the "small for dates" (SFD) baby both perinatally and long-term is discussed, and it is concluded that these children need prompt perinatal care to reduce the

danger of brain damage and death. The SFD baby is defined as any child with a birth-weight below the fifth percentile for gestational age. The neonatal mortality rate for children below the tenth percentile is twice that of the group in the twenty-fifth to seventy-fifth percentile. The usual cause of death is asphyxia. Hypoglycemia is another common disorder of these infants. Several studies suggest a relation between the SFD condition and poor development in later life. Preliminary results of a prospective study of children followed over 5 years suggest that there is a tendency for SFD babies to have subsequent inferior development as gauged by IQ scores. Anticipation of breathing difficulties and hypoglycemia at birth and prompt treatment will lessen the danger of death and brain damage. (22 refs.) - W. Klein.

Department of Child Health
University of Newcastle Upon Tyne
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- 2386 WIGGLESWORTH, J. S. The "small for dates" baby: Pathological and experimental aspects of foetal growth retardation. *Proceedings of the Royal Society of Medicine*, 69(9):879-881, 1967.

The pathological aspects of "small for dates" (SFD) babies which distinguish them from premature and normal infants are discussed, and evidence that the etiology is poor *in utero* maturation is presented. The SFD baby has smaller lungs, liver, and thymus than the premature infant of similar weight, but the brain weight is markedly higher. The decrease in weight in the SFD babies is due to a reduction in cytoplasmic mass, and there is a remarkable similarity between the relative weight and cellular composition of tissue in these babies and others who die of postnatal malnutrition. Thus, an SFD baby may be caused by undernutrition during pregnancy. In an experiment with rats, the growing fetuses were made ischemic, and subsequently, the newborns had many pathologic similarities to SFD babies. In another experiment, a toxin given to pregnant rats late in pregnancy caused growth retardation which seemed to be related to the degree of maternal liver damage (which in turn may have decreased the flow of amino acids to the fetuses). Thus, any condition which reduces the flow of nutrients to a baby may cause the SFD phenomenon. (10 refs.) W. Klein.

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2387 LANGER, LEONARD O., JR. Short stature:

Check list of conditions associated with retarded longitudinal growth. *Clinical Pediatrics*, 8(3):142-153, 1969.

A partial list of conditions associated with short stature is presented for the physician concerned with diagnosis. The categories include: mucopolysaccharidoses and conditions with similar clinical features; bone dysplasias resulting in short limb, disproportionate dwarfism (clinically abnormal at birth); bone dysplasias resulting in short trunk, disproportionate dwarfism; bone dysplasias which may result in asymmetrical limb shortening; bone dysplasias resulting in proportionate dwarfism; primordial dwarfism (low birth-weight dwarfism); chromosomal abnormalities; syndromes which include short stature as a secondary characteristic; and other conditions directly or indirectly responsible for short stature. MR is found in Hurler's disease, Hunter's disease, Sanfilippo's disease, generalized gangliosidosis, Clausen-Dyggve-Melchior syndrome, Albright's hereditary osteodystrophy, Down's syndrome, trisomy 13, penta-X syndrome, *cri-du-chat* syndrome, Brachmann-de Lange syndrome, cerebro-hepato-renal syndrome, Cockayne's syndrome, hypercalcemia, Laurence-Moon-Biedl syndrome, Lowe's syndrome, Menkes' syndrome, Mieten's syndrome, Prader-Willi syndrome, Rubenstein-Taybi syndrome, Sjogren-Larsson syndrome, and the Smith-Lemli-Opitz syndrome. (3 refs.)
M. G. Conant.

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2388 YOUNOSZAI, M. K., & *HAWORTH, J. C.

Chemical composition of the placenta in normal preterm, term, and intrauterine growth-retarded infants. *American Journal of Obstetrics and Gynecology*, 102(2):262-264, 1969.

The chemical composition was determined in blood-free placentas of 26 normal preterm infants (mean birth-weight 2,085 gm), in placentas of 54 normal term infants (mean birth-weight 3,313 gm), and in placentas of 30 term intrauterine growth-retarded infants (mean birth-weight 2,225 gm). The concentrations of water, collagen, and ash were not significantly different in the 3 groups. The placental noncollagen protein concentration of intrauterine growth-retarded infants was significantly higher ($p < .05$) than that of the term and preterm infants who had approximately equal protein concentrations. The preterm infants had significantly higher placental lipid concentrations than the term and

intrauterine growth-retarded infants who had nearly equal concentrations. These chemical substances account for only about 90% of the dry placental weight, and the reported level of glycogen is only 0.5% of the dry weight, thus a large portion is unaccounted for. (12 refs.) - M. G. Conant.

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2389 YOUNOSZAI, M. K., & *HAWORTH, J. C.

Placental dimensions and relations in preterm, term, and growth-retarded infants. *American Journal of Obstetrics and Gynecology*, 103(2):265-271, 1969.

Various measurements were made on placentas (with membranes and cords removed) from 26 normal preterm infants, 85 normal term infants, and 52 term intrauterine growth-retarded infants. The placental weight and decidual area and the cord diameter were significantly less ($p < .001$) in the intrauterine growth-retarded infants than in the other 2 groups while the placental thickness and the fetoplacental weight ratio were significantly less ($p < .05$) in the preterm infants as compared with the other 2 groups. Birth-weight was directly related to placental decidual area in all 3 groups, to placental weight in the term and preterm infants, and indirectly to placental thickness in the intrauterine growth-retarded infants. The placental weight, which reached a constant value by the thirty-sixth week of gestation, was directly related to the placental decidual area which did not increase after the twenty-eighth week of gestation. An important determinant of birth-weight, therefore, may be the decidual area. This, in turn, is likely to be directly related to the chorionic villous surface area. (25 refs.) - M. G. Conant.

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2390 SYBULSKI, S., & TREMBLAY, P. C. Placental glycogen content and utilization in vitro in intrauterine fetal malnutrition. *American Journal of Obstetrics and Gynecology*, 103(2):257-261, 1969.

Glycogen content was measured in 34 placentas from uncomplicated pregnancies, 6 from cases of mild to moderate toxemia, and 13 from pregnancies complicated by intrauterine fetal malnutrition; *in vitro* glycogen utilization was determined by measuring the difference in

glycogen content of the placental tissue before and after incubation. The glycogen content in mg/gm dry tissue was similar in all 3 groups of placentas; however, the total placental glycogen was significantly less ($p < .001$) in the cases of intrauterine fetal malnutrition. Net glycogen utilization was 37% greater than normal in the placentas from cases of intrauterine malnutrition, but this result was not statistically significant. Rates of glycogen utilization of placentas from cases of toxemia were normal. (12 refs.)
M. G. Conant.

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Montreal, Quebec, Canada

2391 KRIEGER, INGEBORG, & WHITTEN, CHARLES F.
Energy metabolism in infants with growth failure due to maternal deprivation, undernutrition, or causes unknown: II. Relationship between nitrogen balance, weight gain, and postprandial excess heat production. *Journal of Pediatrics*, 75(3):374-379, 1969.

Infants with growth failure due to maternal deprivation, malnutrition, or unknown etiology showed an excessive heat production (EHP) postprandially which correlated highly with the rate of weight gain, but appeared to have little relation to protein intake or nitrogen excretion. Sixteen infants and children with no organic disease, but with growth failure related to maternal deprivation or poor food intake, were observed for 3 to 12 days on a diet of 110 calories/kg of body weight, then were given increased feedings to allow rapid weight gain. An average of 146 calories/kg/day led to an average gain of 8.1 gm/kg/day over periods ranging from 23 to 40 days. When feedings were dropped to 124 calories/kg/day, the gain fell to an average of 2.2 gm/kg/day. At the same time EHP, which averaged 45.3% during the period of rapid weight gain, averaged 14.3% during the period of slow weight gain. The correlation between individual EHP and nitrogen retention showed a linear relation during both rapid ($r=.730$) and slow ($r=.807$) weight gain. EHP and weight gain correlated highly ($r=.905$). The relation between EHP and protein or nitrogen intake, however, was poor ($r=.201$). A multiple regression equation was evolved for the total recovery period: $EHP = 2.210 + 0.022 \text{ nitrogen retention} + 4.479 \text{ weight gain}$. It was postulated that EHP related to weight gain is synonymous with EHP related to nitrogen retained for new tissue or that there was inefficient amino acid metabolism secondary to malnutrition. The latter possibility was considered since EHP was at least 100% greater than normal in the Ss, but since the

EHP was associated with rapid weight gain, inefficiency seems unlikely. Studies of normal infants with rapid gain are in process to prove or disprove the postulated causes. (15 refs.) - C. A. Rizvi.

Children's Hospital of Michigan
5224 St. Antoine Street
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2392 BERG, ALAN D. Malnutrition and national development. *Foreign Affairs*, 46(1):126-136, 1967.

Nations are losing valuable mental and physical assets because malnourished children and adults are unable to make their expected contributions. Malnutrition limits life expectancy, decreases a worker's productivity, and lowers the population's resistance to disease. Providing essential nutrients early in life is far less expensive than providing medical costs for treating diseases and other after-effects. Experimental enrichment of rice and flour has already resulted in a decrease in infant mortality in Newfoundland. A new strain of corn was developed with increased lysine which doubles the normal protein value. Lysine may be added synthetically to other foods. Oil seeds (soybean, sesame, sunflower) are being processed into acceptable foods. Fish is another answer, and single-cell organisms growing on petroleum are still another. Since food habits are hard to change, the new nutrient enriched foods must not require a change in buying, cooking, or eating habits. The solution to the problem of malnutrition must be interdisciplinary; however, necessary cooperation has not been achieved in any country. Most governments have not faced up to the magnitude of their role in providing the educational systems which will make the citizens aware of the retarding effects of a malnourished populace. (No refs.) - S. Markworth.

No address

2393 BARNES, RICHARD H. Malnutrition in early life and mental development. *New York State Journal of Medicine*, 65(22):2816-2817, 1965.

Laboratory and human investigations show that malnutrition clearly affects physical and behavioral development, but the relation of malnutrition to MR is not clear. One study, using height as an indicator of malnutrition, found that the shorter children had lower mental test scores. Another study found that as 20 children hospitalized with third degree

protein-calorie malnutrition began to recover their mental scores improved. The Ss from 2.5 to 5 years improved at a greater rate than those from 6 months to 2.5 years, who in turn improved more than the under 6-months-age group. Since the very young infant suffers more from marasmus than kwashiorkor, the relation of these 2 forms of protein-calorie malnutrition should be studied. There is also a need for more studies on the permanence of mental damage. (7 refs.) - S. Markworth.

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2394 EGAN, MARY C. Combating malnutrition through maternal and child health programs. *Children*, 16(2):67-71, 1969.

Public food programs serve only 40% of those who should be served. A Children's Bureau survey found, however, a willingness of state and local agencies to do all they can. Nutritional education must be supported by federal, state, and local governments. Children of low-income families have low dietary intakes of certain vitamins and minerals and low hemoglobin values and as a result they may be stunted in bone development or weight, prone to obesity, and have a propensity to dental caries. Poor nutrition can be caused by lack of money, inadequate storage and cooking facilities, ignorance of good diet composition, parental abuse (intentional withholding of food), or chronic illness. The Children's Bureau supports projects in which nutritionists work with other health and social agencies to program food plans based on individual income, education, and customs. Programed instruction in prenatal education is also being researched. Maternal and child health departments are not only educating parents in how to buy and prepare food, they are demonstrating how sound parent-child relations help children to learn nutrition. Home economists help mothers to read food labels, learn simple housekeeping skills, and buy food wisely. Preschoolers are taught nutrition in day-care and nursery programs. Many community resources are drawn upon to see that parents get not only the education and motivation to change food habits, but the money, food stamps, or donated food to do so. (9 refs.) - S. Markworth.

Children's Bureau
Department of Health, Education, and Welfare
Washington, D. C.

2395 ABELSON, PHILIP H. Malnutrition, learning, and behavior. *Science*, 164(3875): 17, 1969. (Editorial)

Malnutrition has a particularly detrimental effect on learning and behavior and is especially devastating to children under 3 years of age (the period of rapid brain growth). Millions in developing countries and in the United States are experiencing retardation in learning because of inadequate diet. Commercial food products should be enriched, the program of supplying supplementary food to new and expectant mothers should be expanded, and the basic principles of nutrition should be made known to everyone. (3 refs.) - A. Huffer.

No address

2396 MYERS, MADGE L., O'BRIEN, SHEILA CRONIN, MABEL, JUDITH A., & STARE, FREDRICK J. A nutrition study of school children in a depressed urban district. I. Dietary findings. *Journal of the American Dietetic Association*, 53(3):226-233, 1968.

Records of dietary intake over 4 days were kept by 322 fourth-, fifth-, and sixth-graders in 2 schools in a low-income, depressed area of Boston. One-third of the children had 2 or more "unsatisfactory" lunches during this period, 27% unsatisfactory breakfasts, and 14% unsatisfactory evening meals. There were marked deficiencies in the "protective" categories of foods--milk, citrus fruits, and green and yellow vegetables. Negro children had a poorer dietary intake than did Caucasians, and boys had a somewhat poorer intake than girls. The eating pattern of all children seemed to deteriorate at age 10 or 11 years. In order to improve a very real nutritional deficit, it was recommended that boards of education implement school-lunch programs (also a "brunch") and initiate teaching in health, science, and nutrition. (24 refs.) - E. L. Rowan.

Frances Stern Nutrition Center
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2397 MYERS, MADGE L., MABEL, JUDITH A., & STARE, FREDRICK J. A nutrition study of school children in a depressed urban district II. Physical and biochemical findings. *Journal of the American Dietetic Association*, 53(3):234-242, 1968.

Biochemical, anthropometric, and clinical examinations were performed on 322 fourth-, fifth-, and sixth-graders from depressed

urban schools. Many of these children had nutritional deficiencies. Physical examinations revealed a high proportion of dental and gingival pathology, skin lesions, and tongue involvement. Almost 1/2 the children were below the twenty-fifth percentile in height and 1/2 the boys were below the twenty-fifth percentile in weight. Only 8% of the sample was considered obese. About 22% of the sample had hemoglobin values under 14 gm/100 ml. Levels of cholesterol, vitamin A (pooled), carotene (pooled), and thiamine tended to be low. Protein levels as reflected in nitrogen excretion and serum albumin appeared to be low in some Negro children, but the entire group was not examined. Apparent dietary deficiencies, poor hygiene, and lack of fluoridation may contribute to these physical findings. It is significant that no standards exist with which to compare the growth, development, and biochemical findings in urban Negro children. (31 refs.) - E. L. Rouan.

Frances Stern Nutrition Center
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2398 FISHMAN, MARVIN A., PRENSKY, ARTHUR L., & DODGE, PHILIP R. Low content of cerebral lipids in infants suffering from malnutrition. *Nature*, 221(5180):552-553, 1969. (Letter)

Analysis of the cerebral lipids in 4 malnourished infants (2 to 22 mos old) showed that proteolipid proteins, cerebrosides, and plasmalogens--all of which are closely associated with myelin membranes--were consistently reduced. The cerebroside content was 70-80% and plasmalogen content was 80-85% of the control values; proteolipid protein was markedly reduced in the 4-month-old patient, but was not significantly changed in the older cases. Birth-weights in 3 of the 4 cases were lower than the average for infants in the higher socioeconomic section of the same population. Therefore, intrauterine malnutrition and malnutrition during infancy seem inseparable. Although the lipids of myelin class are affected most by malnutrition, it could be an effect secondary to failure to elaborate neural elements or to increase oligodendrocytes. (25 refs.) - L. S. Ho.

Department of Pediatrics and Neurology
Washington University School of Medicine
St. Louis, Missouri 63110

2399 CHASE, H. P., LINDSLEY, W. F. B., JR., & O'BRIEN, DONOUGH. Undernutrition and cerebellar development. *Nature*, 221(5180):554-555, 1969. (Letter)

Undernutrition in young rats significantly decreases the weight and deoxyribonucleic acid (DNA) and protein contents of the cerebellum. There is also a possible decrease of cell numbers. The cerebrum is much less affected; there are less protein and smaller cells in the undernourished cerebrum, but the cell number is not different from control values. The timing of undernutrition is probably important in the extent of final damage. In humans, rapid brain growth commences in the uterus, peaks near birth, and continues during the first year of life. DNA is 2/3 complete at birth in humans. The examination of 2 children who suffered from severe malnutrition in the first 8 months of life and who showed cerebellar damage at age 4 and 5 indicates that postnatal malnutrition is as devastating to humans as it is to rats. (10 refs.) - L. S. Ho.

University of Colorado Medical Center
Denver, Colorado 80220

2400 BIRCH, HERBERT G. Problems inherent in population studies of nutrition and mental subnormality. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 8, p. 57-62.

To analyze the relation between malnutrition in childhood and mental or intellectual dysfunction, the biological attributes associated with poverty, the family environment, and individual psychological experiences must be considered. Evaluation of the effects of biological variables should include data on sibships, the nutritional status of the mother as a child, and the exaggerated effects of infectious disease on malnourished children as well as longitudinal data on the pregnancy, delivery, and neonatal characteristics of nutritional risk children. Since the organization of the family environment of malnourished children may be different from that of well nourished children, the differential factors involved in the ecology of growth and their relation to malnutrition must be identified. Consequences of malnutrition which modify individual psychological development and experiences include: behavioral unresponsiveness; failure to learn during critical periods; initial lag in mental development; and reduced parent-child interaction. Longitudinal studies of at-risk populations which control for social, ecological, and biological factors are needed. (No refs.) - J. Wyatt.

2401 CRAVIOTO, JOAQUIN. Protein-caloric undernutrition and mental retardation. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 9, p. 63-76.

Severely malnourished infants (CA 6-42 mos) evidenced impaired performance in adaptive, motor, language, and social behavior. Developmental quotients of most Ss increased as recovery from malnutrition occurred; however, infants who suffered from malnutrition prior to 6 months of age had persistently low scores in adaptive behavior during rehabilitation. Malnutrition appears to increase the chances of a below normal score on an intelligence test and reduces the chances of a superior score. A potential permanent reduction in intellectual potential and delayed development in intersensory functioning appear to occur in infants subjected to malnutrition at a very early age. Malnutrition may affect mental development by reducing the amount of learning time, interfering with learning during critical periods, and reducing motivation. A child with inadequate intersensory development during the preschool years may fail to establish an ordinary normal background of conditioning and may then fail to profit from educational exposure during the school years. (37 refs.)

J. K. Wyatt.

2402 DOBBING, JOHN. The effects of experimental undernutrition on the developing brain. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 10, p. 77-86.

The effects of undernutrition on the developing brain and the persistence of these effects into later life appear to be related to the severity and duration of the undernutrition as well as to timing in relation to the time of the maximum period of brain growth. A large number of brain constituents have a degree of metabolic stability comparable to that of relatively inert tissues. When these constituents are laid down during a period of maximum brain growth, they remain relatively stable throughout life. It may be that when the myelin constituents are being actively synthesized within the brain, the brain is particularly susceptible to the effects of undernutrition and that even comparably small restrictions at this time can have large and permanent effects. Even when undernutrition in guinea pigs did not begin until the period of maximum brain growth was almost completed, there was permanent retardation

and full rehabilitation could not be achieved. Rats undernourished from birth evidenced reduced body and brain weight during the second and third weeks of life as well as considerably reduced brain cholesterol content. However, the brains of rats undernourished from 3 to 4 weeks of age to 11 weeks of age had a higher cholesterol concentration than those of their normal controls. (12 refs.)

J. K. Wyatt.

2403 DODGE, PHILIP R. Nutrition and the developing nervous system. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 11, 87-89.

The type of malnutrition which affects the greatest number of infants and children in the world and constitutes the bulk of the problem in the field of nutrition is protein-calorie undernutrition. The majority of children throughout the world are fed diets which are deficient in both calories and protein. Undernutrition in early life has been related to significantly reduced head circumference for age and initial intelligence quotients below 90. Additional research is needed to determine: the effects of nutrition on growth, maturation, and function of the developing nervous system; the relation between undernutrition during infancy and level of intelligence reached at adulthood; and the role of undernutrition in the etiology of MR. (9 refs.) - J. K. Wyatt.

2404 DE SILVA, C. C., & BAPTIST, N. G. *Tropical Nutritional Disorders of Infants and Children*. Springfield, Illinois, Charles C. Thomas, 1969, 226 p. \$11.50.

Aspects of deficiencies of carbohydrates, lipids, proteins, vitamins, and minerals among children in tropical countries were discussed. In particular, the interrelation of calories and proteins in the various types of kwashiorkor and marasmus were studied. Genotypical and environmental factors affecting growth were discussed as well as erythropoiesis and anemia. Physicians, clinicians, and public health officials should profit from perusal of this book. (713 refs.)

L. S. Ho.

CONTENTS: Some General Considerations; Growth Failure of Growth; Carbohydrates; Lipids; Proteins; Vitamins; Minerals; Erythropoiesis and Anemia.

2405 WINICK, MYRON, & ROSSO, PEDRO. Head circumference and cellular growth of the brain in normal and marasmic children. *Journal of Pediatrics*, 74(5):774-778, 1969.

Head circumference was found to be a good measure of cellular brain growth in normal and malnourished infants. The normal population consisted of 3 fetuses and 7 children from Santiago (Chile) who had died from accidents, poisonings, or therapeutic abortions. The head circumference was measured in all of these children just prior to autopsy. The total brain weight and total brain protein increased more rapidly than head circumference during the first year of life. The brain deoxyribonucleic acid (DNA) content was linearly related to head circumference during the first years of life. Equations for total brain weight to head circumference, total brain protein to head circumference, and total brain DNA to head circumference are given. From these equations, the total brain weight, protein, and DNA can be calculated for the normal child for any given head circumference. All the malnourished children had head circumferences more than 2 standard deviations below the mean for normal children whose ages were less than 1 month up to 16 months. The brain weight and protein were reduced proportionally to the reduction in head circumference. DNA content or cell number was reduced at least as much as, and in 3 cases, more than, head circumference. The degree of reduction of head circumference in children malnourished during the first year of life, therefore, indicates the severity of the nutritional deprivation on the actual cellular growth of the brain. (7 refs.)

F. J. McNulty.

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2406 WHARTON, BRIAN, HOWELLS, GLAN, & PHILLIPS, IAN. Diarrhoea in kwashiorkor. *British Medical Journal*, 4(5631):608-611, 1968.

Seventy-one children with kwashiorkor admitted to a malnutrition research unit in a Kampala hospital were treated with a combined chemical, biochemical, and bacteriological approach. Sugar intolerance was noted as a common cause of diarrhea in the children. A lactose-free diet reduced the sugar intolerance when there was no enteric infection. If the child was still intolerant, fructose was substituted for the sucrose. Antibiotics were of limited value. In the stool examination, 14 children had severe diarrhea, 30 children had moderate diarrhea, and 27 children had mild diarrhea. A specific enteric infection was found in 6 children, and all but one of these had severe diarrhea; 5 of these 6 children had septicemia with intestinal sugar intolerance; *Salmonella staley* was found in one child and *S. garoli* was found in the other 5. The 6 children and an additional 15 children had sugar in their stools. The diarrhea was moderate in 8 and severe in 5 of these children. Lactose intolerance predominated in 21 children with sugar intolerance and persisted in 3 children for some weeks. Intolerance to monosaccharides and disaccharides was less common and persisted in one child. Sugar loads were determined in 16 intolerant children, and of the 6 children with enteric pathogens in the stool, 3 had mixed intolerance to various sugars, excluding fructose, and 3 were intolerant of all sugar including fructose. (16 refs.) - F. J. McNulty.

Infantile Malnutrition Research Unit
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New Growths

- 2407 RIZZUTO, N., & FERRARI, G. Familial infantile myoclonic epilepsy in a family suffering from tuberous sclerosis. *Epilepsia*, 9(2):117-125, 1968.

A study of a mother and 4 children, all of whom experienced cutaneous and cerebral symptoms of tuberous sclerosis and epileptic phenomena with massive spasms, links tuberous sclerosis with infantile myoclonic epilepsy hypsarrhythmia (IMEH). The mother (CA 42 yrs) who was MR, had skull calcifications at the basal ganglia, vertex, and the ventricular trigone. Her EEG was characterized by a rhythm of 9-10c/sec, and rhythmic spike waves were recorded. The oldest female (CA 12 yrs) showed butterfly sebaceous adenomas on the forehead, cheeks, nose, and chin. There was no record of convulsive seizure, but MR was severe. Calcifications were also present in her skull, and her EEG showed a well organized bilateral, symmetrical rhythm of 10c/sec with isolated bilateral spike and waves. A son (CA 8 yrs) presented massive spasms at 4 months, and his mental development was arrested at this age. At 2 years, Pringle adenomas appeared. Calcifications were present in the basal ganglia area, and his EEG indicated a regular alpha rhythm of 10 c/sec. A daughter (CA 4 yrs) developed flexion spasms and loss of psychomotor functions at 4 months. No mental defect was disclosed, and calcifications were found close to the ventricular surface. The second son (CA 3 yrs) was SMR. He developed flexion spasms at 4 months, and adenomas appeared at 2 years. His EEG showed typical hypsarrhythmic features, and calcifications were disclosed near the coronal suture. The 3 younger children exhibited West's syndromes in early childhood. The importance of these observations is that: tuberous sclerosis is important in the etiology of IMEH; a familial form of IMEH exists which heralds tuberous sclerosis; and no direct correlation exists between IMEH and MR. (17 refs.) - V. G. Votano.

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Nervose e Mentali dell' Università
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Cagliari, Italy

- 2408 CHANDOR, S. B., & OSTERAAS, G. R. Congenital glioma in a stillborn infant. *Military Medicine*, 134(4):257-258, 1969.

A 7-pound infant delivered stillborn at term showed hydrocephalus and a glioma of the left cerebral hemisphere and represented the tenth reported case of congenital glioma. Although the pregnancy had been uneventful, hydrocephalus was discovered at delivery. At autopsy, pathology was confined to the head and brain. The occipital circumference was 40 cm, the fixed brain weighed 500 gm, and a firm, tan-white tumor was found occupying the left cerebral hemisphere, pons, and medulla. Microscopic examination revealed a low-grade glioma which appeared to arise from the left cerebral hemisphere; however, exact placement of origin was not possible. More frequent postmortem examinations should increase the known cases of this rare tumor. (4 refs.) W. Klein.

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University of Southern California School
of Medicine
Los Angeles, California 90033

- 2409 DURITY, F. A., DOLMAN, C. L., & MOYES, P. D. Ganglioneuroblastoma of the cerebellum: Case report. *Journal of Neurosurgery*, 28(3):270-273, 1968.

A 3-year-old female who had a history of lethargy, vomiting, and occipital headaches, underwent suboccipital craniotomy for removal of an encapsulated tumor in the right cerebellar hemisphere. Her coordination showed continuing improvement postoperatively. The tumor contained a mature and an immature element which existed in microscopically distinct areas as well as intermediate forms between the neuroblasts and the mature neuroones. This ganglioneuroblastoma is probably a transitional phase in the evolution of a neuroblastoma to a gangliocytoma and is the first reported case in a cerebellar hemisphere. Only 2 of the 7 patients reported

received surgery and they survived for 5-6 years, so the prognosis is not entirely hopeless. (19 refs.) - M. G. Conant.

Vancouver General Hospital
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2410 DASTUR, D. K., & LALITHA, V. S. Pathological analysis of intracranial space-occupying lesions in 1,000 cases including children: Part 2. Incidence, types and unusual cases of glioma. *Journal of the Neurological Sciences*, 8(1):143-170, 1969.

Of the 1,000 intracranial space-occupying lesions analyzed, 370 were gliomas which occurred in patients with an average age of 27 years. Children under the age of 15 accounted for 27.3% of all gliomas, two-thirds of which was infratentorial. Types of glioma observed were, in order of decreasing incidence, astrocytomas, ependymomas, medulloblastomas, optic nerve gliomas, neurohypophyseal gliomas, pineal tumors, choroid plexus papilloma, ganglioglioma, medulloepithelioma, olfactory neuroblastoma, and cerebral reticulosis. Astrocytomas comprise 60.5% of all gliomas; 39.3% of these are benign and 60.7% are malignant. The average age for Ss with astrocytomas is 31.5 years. (41 refs.) M. G. Conant.

J. J. Group of Hospitals
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2411 GROVER, WARREN D., & *RORKE, LUCY BAILIAN. Invasive craniopharyngioma. *Journal of Neurology, Neurosurgery, and Psychiatry*, 31(6):580-582, 1968.

A 17-year-old boy succumbed to a craniopharyngioma which had been partially removed when he was 5 and irradiated when he was 14. Direct invasion by a craniopharyngioma is rare but was diagnosed in this case by progressive focal neurological signs. Initially, the patient developed headache, vomiting, visual field defects, and ataxia. Progressive invasion was documented by the appearance of dysarthria, dysgraphaesthesia and astereognosis, hemiparesis, aphasia, and dysphagia. At autopsy, there was extensive invasion and destruction of the caudate, thalamus, midbrain and pons. (2 refs.) - E. L. Rowan.

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Philadelphia, Pennsylvania 19146

2412 CANTU, ROBERT C., ADAMS, RAYMOND D., McNAMARA, JOHN, & *CONNELLY, JOHN P. Spinal cord tumors in children. *Clinical Pediatrics*, 7(12):726-732, 1968.

An 8-year-old boy with a history of pain and spasm of the back muscles was found to have a spinal cord tumor which was demonstrated by myelography and was surgically extirpated. Keratin spillage associated with this epidermoid tumor resulted in an aseptic meningeal reaction which responded quickly to glucocorticoid medication. Excision is curative, and radiotherapy and chemotherapy have no place in the treatment of epidermoid tumors although these latter methods are the treatment of choice in inoperable gliomas, malignant epidural or subdural growths, and tumors of the neuroblastic type. It is essential that the clinician be alert to the manifestations of spinal cord tumors (local pain or mass and root symptoms) because of the potential curability following early intervention. (5 refs.) - E. L. Rowan.

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2413 MOYES, PETER D. Intracranial and intraspinal vascular anomalies in children. *Journal of Neurosurgery*, 31(3):271-278, 1969.

Case histories of 13 children with intracranial or intraspinal vascular anomalies indicated that in all but one S, symptoms became apparent as a result of hemorrhage. Only 2 of these patients had seizures; this is not consistent with studies on adults with similar anomalies. Mortality and morbidity figures for these 13 children are considered low, and in 10 cases there was complete removal of the lesion. Seven of the 13 patients had lesions in the posterior head region or spinal canal. Posterior circulation angiography is cited as essential in children with spontaneous intracranial hemorrhages when anterior circulation angiography does not indicate the reason for the hemorrhage. Data for the 13 cases show 1 death, 4 children with mild neurological deficits, and 8 with no deficits. (24 refs.) - B. Bradley.

University of British Columbia
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- 2414 MOYES, PETER D. Subdural effusions in infants. *Canadian Medical Association Journal*, 100(5):231-234, 1969.

Among 60 children with documented subdural hematomas in infancy, the most common causes were trauma (birth, accidental, or "battering") or as a sequel to meningitis. The slow collection of fluid produced few specific signs, but rather caused irritability, psychomotor retardation, and failure to thrive. Treatment was directed at evacuation of the effusion and correction of nutritional deficiencies. Initially, a subdural tap was done through the anterior fontanelle or coronal suture. If necessary, burr holes were placed for more extensive drainage and shunting was done for approximately 6 weeks. Only infrequently were membranes removed surgically. In this series there were 47 good results, 10 patients with residual deficits, and 3 deaths. Treatment is successful if instituted soon after the insult; therefore, a high index of suspicion of subdural effusion is necessary when evaluating the irritable infant. (13 refs.) - E. L. Rowan.

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- 2415 MORAGAS, A., & VIDAL, MARIA-TERESA. Giant congenital intracranial teratoma. *Helvetica Paediatrica Acta*, 24(1):106-110, 1969.

A giant cerebral teratoma was the cause of macrocephaly in a female infant (weight 2,480gm) who died 30 minutes after cesarean birth. External data included a cranial perimeter of 47 centimeters, a parchment-like skull, and a wide fontanelle. Macroscopic examination revealed normal meninges which enclosed a lobulated mass (weight 501 gm) that had no discernible corticocerebral textures but contained cavities of citrine liquid and centers of pebbly calcification, an immature cerebellum, flat sella turcica and skull floor, and normal eyeballs and pituitary gland. Microscopic examination of the cerebral mass showed neuroepithelial structures, glial plates, ependymal-like formations, choroidal plexus structures, optical vesicles, cartilaginous-bony areas, and epithelium-lined glandular spaces. The diagnosis of macrocephaly, as opposed to hydrocephaly, was made possible by the postmortem examination. (9 refs.) - A. Huffer.

Children's Hospital
Barcelona, Spain

- 2416 LEE, DANIEL K., & ABBOTT, MICHAEL L. Familial central nervous system neoplasia: Case report of a family with von Recklinghausen's neurofibromatosis. *Archives of Neurology*, 20(2):154-160, 1969.

The combination of posterior polar cataracts with central nervous system (CNS) neoplasia in a family is reported for the first time. Eight of 17 family members were affected with CNS tumors. Only minor evidence of peripheral von Recklinghausen's disease exists in this family. Three patients had peripheral neurofibromas, and no patient had more than 2 cafe-au-lait spots. One patient with a proven neurofibroma was considered to have von Recklinghausen's disease. The multiple CNS tumors in association with von Recklinghausen's disease suggest that these 2 entities are related. In this family, 3 patients had bilateral eighth nerve neurinomas associated with multiple meningiomas and one associated with hypothalamic astrocytoma. A single patient had bilateral eighth nerve neurinoma and one had parasagittal meningioma. Seven of the 17 patients had congenital ocular abnormalities. Congenital squint was present in one of these, and either unilateral or bilateral posterior lenticular opacities and squint were present in the remaining 6. (22 refs.) - F. J. McNulty.

Department of Medicine
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- 2417 FRAUMENI, JOSEPH F., & MILLER, ROBERT W. Cancer deaths in the newborn. *American Journal of Diseases of Children*, 117(2):186-189, 1969.

Review of United States national mortality figures for 1960-1964 revealed that 130 children died of cancer within the first 28 days of life. Leukemia (44) and neuroblastoma (27) accounted for more than half the deaths, but teratoma (9) and primary liver cancer (10) showed relatively high frequencies. There were no sex differences for neonatal cancers even though boys are more susceptible to neoplasia later in childhood. No significant temporal, geographical, or sibling aggregational factors were found. Congenital defects were reported on 12 of the death certificates. Eight of the 44 leukemia cases had Down's syndrome. This was the only finding consistent with the hypothesis that defective prenatal development is associated with a

higher frequency of neonatal neoplasia. (17 refs.) - E. L. Rowan.

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2418 FRAUMENI, JOSEPH F., JR. The aniridia-Wilms' tumor syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original

Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 198-201.

The syndrome of congenital aniridia and Wilms tumor has now been recorded in 24 patients. Associated defects include MR, genitourinary tract anomalies, retarded growth and development, microcephaly, and anomalies of the external ear. All but one case have been sporadic, and the origin of the syndrome is likely to be either a fresh gene mutation or an environmental effect on embryogenesis. The role of abnormal growth mechanisms in oncogenesis is uncertain. (15 refs.)
E. L. Rowan.

Prenatal Influence

2419 CONDRON, COLIN J. Limb anomalies in the Cornelia de Lange syndrome--Infant patient. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 226-227.

An infant girl with peculiar facies, hirsutism, hoarse cry, and MR characteristics of the de Lange syndrome also showed minor limb anomalies. Her hands and feet were disproportionately small, the thumbs were proximally placed, the fifth fingers showed microclinodactyly, and there was partial pedal syndactyly. (No refs.) - E. L. Rowan.

2420 FILIPPI, GIORGIO, & RENUART, A. W. Limb anomalies in the Cornelia de Lange syndrome--Adult patient. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 228-229.

An SMR 47-year-old female with clinical features of the de Lange syndrome was also found to have limb anomalies including a lobster-claw deformity of the right hand, a simian

line and clinodactyly of the left hand, and brachydactyly of the toes. (No refs.)
E. L. Rowan.

2421 SMITH, G. F., BERG, J. M., & McCREARY, B. D. De Lange syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 18-21.

The de Lange syndrome has been recorded in over 200 patients, but unfortunately, there is no pathognomonic clinical manifestation. Generally, there is a birth-weight less than 2,500 gm, MR, growth retardation, and hirsutism. Characteristic facies show microcephaly, confluent eyebrows, long eyelashes, small nose with anteverted nostrils, increased distance from nostrils to lip, and micrognathia. Common limb findings include small hands and feet, proximally-placed thumbs, and absent or deformed bones of the upper limbs. No consistent chromosomal abnormality has been found and no pattern of inheritance uncovered. Because of the nonspecific picture more than one clinical entity may be involved or more than one causal factor may lead to the same clinical findings. (8 refs.)
E. L. Rowan.

2422 RUBINSTEIN, JACK H. The broad thumbs syndrome--Progress report 1968. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 25-41.

A total of 114 cases of the "broad thumbs" or Rubinstein-Taybi syndrome have been collected since it was first described in 1963. The most characteristic clinical findings include MR, motor, language, and social retardation, broad terminal phalanges of thumbs and great toes, short stature, head circumference and bone age below the 50th percentile, beaked or straight nose, high-arched palate, antimongoloid slant to palpebral fissures, and delayed or incomplete descent of testes in males. Many other anomalies may accompany the syndrome for which no etiology has yet been determined. Case finding must continue, and patients and families should be studied extensively to determine the true parameters, natural history, and cause(s) of this syndrome. (47 refs.) - E. L. Rowan.

2423 ROBINOW, MEINHARD. A familial syndrome of mental deficiency and broad thumbs. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 42.

A mother and her 4 children were all affected with MR, distinctive narrow facies with prominent noses and antimongoloid slant to the eyes, broad thumbs and toes, and feeding difficulty at birth. Although these features are suggestive of the Rubinstein-Taybi syndrome, the patients were uniformly tall, MR was mild, no cryptorchism was noted, dermatoglyphics were unremarkable, and there were no other associated congenital abnormalities. This family, then, appears to have a distinct syndrome which might be mistaken for the Rubinstein-Taybi syndrome. (No refs.) E. L. Rowan.

2424 DIEKER, H., EDWARDS, R. H., ZURHEIN, G., CHOU, S. M., HARTMAN, H. A., & OPITZ, J. M. The lissencephaly syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 53-64.

The central nervous system manifestations of the lissencephaly syndrome (smooth brain surface, internal hydrocephalus, and disturbed

gray matter) are apparent only at autopsy, but clinical manifestations are such that the syndrome can be diagnosed during life. Infants present with severe psychomotor retardation, height and weight below the third percentile, microcephaly, micrognathia, hirsutism, a characteristic combination of minor facial anomalies, minor hand anomalies, frequent congenital heart disease, and hepatosplenomegaly. Hypotonia progresses to decerebrate posturing, and such patients rarely survive infancy. Familial occurrence suggests an autosomal recessive mode of inheritance. (18 refs.) - E. L. Rowan.

2425 GORLIN, ROBERT J. Some facial syndromes. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 65-76.

There are at least 8 specific malformation syndromes which affect the face. The popliteal pterygium syndrome also includes genital hypoplasia and cleft lip-palate. The multiple mucosal neuroma syndrome consists of nodules on any or all mucosal surfaces and is inherited as an autosomal dominant trait. The Charlie M. syndrome consists of facial paralysis, conical incisors, hypertelorism, and digital anomalies. Frontonasal dysplasia or midfacial cleft syndromes include the wide range of patients with hypertelorism. Whistling face syndrome derives its name from the characteristic tiny mouth. Oculo-dento-digital syndrome includes microcornea, hypoplasia of alae nasi, absent tooth enamel, and 4-5 syndactyly. Frontometaphyseal dysplasia is manifest as a torus-like structure extending from eye to eye and anomalous supra-orbital bone. Oral-facial-digital syndromes of Type I are lethal to males while females show a pseudo-cleft of the upper lip, lobulated tongue, mild MR, replication of teeth, and a wide spectrum of hand anomalies. (No refs.) - E. L. Rowan.

2426 BIXLER, DAVID, CHRISTIAN, JOE C., & GORLIN, ROBERT J. Hypertelorism, microtia and facial clefting: A new inherited syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 77-81.

Two sisters (CA 8 and 12 yrs) have a similar malformation syndrome of hypertelorism, microtia, and clefting of the lip, palate, and nose. Both are considered to be MR, but

atretic auditory canals with resultant hearing loss make it difficult to assess intelligence. The girls also have microcephaly, steep mandibular angles, ectopic kidneys, and congenital heart disease. There is a strong history of congenital heart disease on the mother's side of the family. This new syndrome must be differentiated from the median cleft face syndrome and the oro-palato-digital syndrome and is probably inherited as an autosomal recessive trait. (5 refs.)

E. L. Rowan.

- 2427 CHRISTIAN, JOE C., BIXLER, DAVID, BLYTHE, SUE C., & MERRITT, A. DONALD. Familial telecanthus with associated congenital anomalies. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 82-85.

Telecanthus (distance between medial canthi greater than 2 standard deviations above the mean) was noted in 8 members of a family over 5 generations. Besides telecanthus, the proband showed cranial asymmetry, widow's peak, cleft lip and palate, strabismus, imperforate anus, hypospadias, and flame nevi. Ureteral stenosis and cryptorchidism had also been noted in a brother of the proband who died in infancy and had the same malformation syndrome. Telecanthus appeared to be a dominant family trait, and no instance of male-to-male transmission was observed. The associated congenital anomalies are postulated to be manifestations of a single mutant gene. Although MR was not a part of the syndrome in this family, it was prominent in another reported family with a similar constellation of physical malformations. (10 refs.) - E. L. Rowan.

- 2428 OPITZ, JOHN M., SUMMITT, ROBERT L., & SMITH, DAVID W. The BBB syndrome--Familial telecanthus with associated congenital anomalies. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 86-94.

Nine males in 3 separate families manifested hypertelorism and variable degrees of hypospadias. MR, cleft palate, cryptorchidism, and congenital heart disease were also noted in some of the affected men. The condition

appears to be inherited as an autosomal dominant trait with severe manifestations appearing only in males. Carrier women do appear to have telecanthus. The penile deformity is probably a defect of the genital tubercle. Since there were 3 sets of twins in 2 of these families, there may be an etiologic relation between twinning and this syndrome. (10 refs.) - E. L. Rowan.

- 2429 OPITZ, JOHN M., FRIAS, JAIME L., GUTENBERGER, JAMES E., & PELLETT, JOHN R. The G syndrome of multiple congenital anomalies. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 95-101.

A previously undocumented malformation syndrome occurred in all 4 brothers in the "G" family. The most significant manifestation was neuromuscular dysfunction of the esophagus with choking and cyanosis on feeding and failure to thrive. Three of the brothers died in infancy, and the survivor was fed via a gastrostomy. Each had a hoarse cry, prominent parietal eminences, occipital prominence, hypertelorism, slanting ears, and hypospadias. The mother and one sister reportedly had swallowing difficulty in infancy, and the mother also had hypertelorism and slanting ears. Etiology of this syndrome is probably genetic with incompletely recessive X-linked inheritance or autosomal dominant inheritance with male sex limitation and partial female heterozygous expression the most likely mechanism. (4 refs.) - E. L. Rowan.

- 2430 THULINE, H. C. Current status of a family previously reported with the oral-facial-digital syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 102-103.

Follow-up of the pregnancy history of a family with the oral-facial-digital (OFD) syndrome reveals a total of 49 pregnancies in affected women. There were 17 affected and 9 unaffected females, 1 male with *forme fruste* and 14 unaffected males born. The syndrome is widely believed to be lethal to males, and if the 8 spontaneous abortions are

considered to be male, then this belief is supported by a reasonable overall sex ratio of male to female conceptions of 1:1.1. Unilateral polydactyly was found in 5 of the affected females, thus confirming an earlier observation that this is a pleomorphic manifestation of the lethal-to-male Type I of the OFD syndrome. (3 refs.) - E. L. Rowan.

2431 OPITZ, JOHN M., & FAITH, GLENN C. Visceral anomalies in an infant with the Goldenhar syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 104-105.

The Goldenhar syndrome is not confined to malformations of the "first arch" and vertebral column, with or without epibulbar dermoids, but is a complex dysmorphogenetic syndrome with multiorgan involvement. Rectal prolapse, coarctation of the aorta, unilobular lungs, and hemosiderosis of the liver were noted in one patient and other cardiac, pulmonary, renal, gastrointestinal and central nervous system malformations have been described. The etiology of this syndrome is unknown. (5 refs.) - E. L. Rowan.

2432 SUMMITT, ROBERT. Familial Goldenhar syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 106-109.

An infant had several manifestations of the Goldenhar syndrome including an epibulbar dermoid, malformed auricles and preauricular appendages, malar hypoplasia, prominent micrognathia, and cleft palate. Pedigree analysis showed the father to have preauricular appendages, micrognathia, and low normal intelligence. Less severe forms of the same condition were found in 7 other relatives over 3 generations. The syndrome appears to be inherited as an autosomal dominant trait, but the wide variability of expressivity and the presence of an epibulbar dermoid only in the proband raises the possibility that oculoauriculovertebral dysplasia (Goldenhar syndrome) and the similar syndromes of mandibulofacial dysostosis and hemifacial microsomia may be variations of the same mutant gene. (5 refs.) - E. L. Rowan.

2433 HERRMANN, JURGEN, & OPITZ, JOHN M. A dominantly inherited first arch syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 110-112.

At least 14 members of a family are affected with a first arch malformation syndrome ranging from minor anomalies to a full-blown syndrome of a transverse oral cleft and preauricular appendages in one proband and occipital encephalomeningocele, hydrocephalus, MR, preauricular appendages, and micrognathia in a second. The inheritance pattern is best explained by autosomal dominant transmission with incomplete penetrance. The wide variability found in this family suggests that syndromes based on sporadic cases of first arch anomalies may represent invalid splitting. (9 refs.) - E. L. Rowan.

2434 COHEN, M. MICHAEL, & GORLIN, ROBERT J. Genetic considerations in a sibship of cyclopia and clefts. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 113-118.

A 14-member sibship includes 4 children with cleft lip and/or palate, 1 with bifid uvula, and 1 with cyclopia. The parents are consanguineous Chippewa Indians. This is the second reported family in which a holoprosencephalic disorder (cyclopia, ethmocephaly, clebocephaly, and premaxillary agenesis) has been associated with clefts. Although this cyclops had a normal karyotype, various chromosomal anomalies have been reported in other cases. No other abnormalities were noted in the children with clefts. The genetic basis of human holoprosencephaly has not been determined, and more case reports with extensive investigations of probands and siblings are indicated. (86 refs.) - E. L. Rowan.

2435 OPITZ, JOHN M. Familial anomalies in the Pierre Robin syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 119.

The Pierre Robin syndrome is considered to arise sporadically, but 2 families have been

noted in which anomalies have occurred in relatives. Both probands had micrognathia, retroglossia, and cleft palate, and one had the commonly associated findings of ophthalmic malformations and MR. In one family, a sister of the proband had a cleft palate, and in the other, both the mother and sister of the proband had a cleft palate and cataracts. A genetic mechanism is suggested, but the mode of the transmission is yet to be determined. (1 ref.) - E. L. Rowan.

2436 FRASER, G. R. Malformation syndromes with eye or ear involvement. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 130-137.

Malformation syndromes are frequently seen among deaf and blind children, and the sensory impairments may represent pleiotropic effects of a single gene defect. Both the eye and ear are involved in the autosomal recessive cryptophthalmos syndrome. Retinal lesions with microcephaly and cataracts with chondrodystrophia calcificans are also autosomal recessive syndromes, whereas dominant forms of inheritance are found in the Hallermann-Streiff syndrome, blindness with skeletal dysplasias, and the Pierre Robin syndrome. Deafness is associated with dominantly inherited Treacher Collins syndrome, branchial arch syndrome, and the Waardenburg syndrome. Deafness with split hand and foot is an autosomal recessive condition. Both deafness and blindness are components of rubella embryopathy, so that both genetic and environmental insults may result in complex birth defects. Attempts to identify the etiology of these defects may aid in unraveling the mysteries of growth and development. (42 refs.) - E. L. Rowan.

2437 HERRMANN, JURGEN, & OPITZ, JOHN M. The Lenz microphthalmia syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 138-143.

The Lenz microphthalmia syndrome was observed in an 11-year-old boy with skeletal abnormalities, microphthalmia, renal dysgenesis, dental anomalies, hypospadias, speech impairment, and psychomotor retardation. An X-linked recessive mode of inheritance has been

suggested, and minor abnormalities were seen in the proband's mother (short stature, microcephaly, syndactyly, and unusual dermatoglyphics) and brother and sister (syndactyly and unusual dermatoglyphics). (5 refs.)

E. L. Rowan.

2438 OPITZ, JOHN M., ZURHEIN, GABRIELLE M., VITALE, LEONARD, SHAHIDI, NASROLLAH T., HOWE, JOHN J., CHOU, SHI MING, SHANKLIN, DOUGLAS R., SYBERS, HARLEY D., DOOD, ARNOLD R., & GERRITSEN, THEO. The Zellweger syndrome (Cerebro-hepato-renal syndrome). In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 144-158.

Extensive pathological and histochemical studies of 4 additional cases (total of 13) of the cerebro-hepato-renal syndrome of Zellweger revealed other anomalies besides severe congenital hypotonia, profound psychomotor retardation, characteristic facies, and the minor anomalies of the extremities usually associated with this syndrome. All patients had small, micromulticystic kidneys. Livers were enlarged, diffusely fibrotic, and contained deposits of hemosiderin. The etiology of this abnormal iron storage is unclear. Neuropathological examination revealed olfactory agenesis, hydrocephalus, micropolygyria, pachygyria, cerebral edema, and a myelin deficiency considered to represent a leukoencephalomyelopathy. The Zellweger syndrome is probably an autosomal recessive disorder. (13 refs.) - E. L. Rowan.

2439 TAYLOR, JAMES C., ZELLWEGER, HANS, & HANSON, JAMES W. A new case of the Zellweger syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 159-160.

A male infant with the same constellation of anomalies was born to the family in which the Zellweger syndrome was first described in 1964. Like his sibling, the patient had marked hypotonia, characteristic facies, hepatosplenomegaly, dermal ridge hypoplasia, and increased serum iron. The clinical course was characterized by apneic spells and seizures. Gross pathological examination revealed multicystic kidneys and cardiac anomalies. (2 refs.) - E. L. Rowan.

2440 OPITZ, JOHN M., JOHNSON, RONALD C., McCREADIE, SAMUEL R., & SMITH, DAVID W. The C syndrome of multiple congenital anomalies. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 161-166.

Two siblings of the "C" family manifested the same dysmorphogenetic syndrome and died in infancy with jaundice and failure to thrive. Anomalies included: accessory digits; joint deformities; skull, face, and ear anomalies; laxity of the skin; syndactyly; and short stature. Autopsies revealed hepatomegaly, patent ductus arteriosus, and immaturity of kidneys and lungs. The anterior fossa of the skull was hypoplastic, there was an osseous defect between the orbits, and the tentorium was incompletely developed. This syndrome is different from the Ellis-van Creveld syndrome and is probably inherited as a recessive mutation. (1 ref.) - E. L. Rowan.

2441 OPITZ, JOHN M., & HOWE, JOHN J. The Meckel syndrome (Dysencephalia splanchnocystica, the Gruber syndrome). In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 167-179.

Sloping forehead, posterior exencephalocele, polydactyly, and polycystic kidneys are the most striking findings in a dysmorphogenetic syndrome first described by Meckel in 1822 and reintroduced in the modern literature by Gruber in 1934. Only 4 cases have been recorded in the English language and the syndrome is generally unknown. The Meckel syndrome is apparently recessively inherited and lethal in the perinatal period. Associated anomalies of the skull, brain, eyes, ears, limbs, genitalia, lungs and kidneys have been recorded. The so-called Ullrich-Feichtiger syndrome probably includes patients with D₁ trisomy, Smith-Lemli-Opitz syndrome, and milder forms of the Meckel syndrome. (32 refs.) - E. L. Rowan.

2442 BECKWITH, J. BRUCE. Macroglossia, omphalocele, adrenal cytomegaly, gigantism, and hyperplastic visceromegaly. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects

Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 188-196.

A syndrome characterized by macroglossia, omphalocele, and renal hyperplasia can be diagnosed at birth. Other common features are cytomegaly of the adrenal cortex, pancreatic and gonadal interstitial cell hyperplasia, and peculiar facies. Neonatal hypoglycemia occurs frequently and is the most probable cause of early death. Survivors show postnatal gigantism, occasional hemihypertrophy, a high risk of MR, and an increased frequency of intraabdominal neoplasia. Much work needs to be done in this apparently autosomal recessive syndrome to understand the interrelations of the hyperplastic process and endocrine dysfunction. (23 refs.) - E. L. Rowan.

2443 COHEN, M. MICHAEL, JR. Comment on the macroglossia-omphalocele syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 197.

Macroglossia appears to be a constant, but variable, feature of the macroglossia-omphalocele syndrome, and longitudinal study is essential to an understanding of the growth and development of the dentofacial complex. The frequency of malocclusion, prognathism, and anterior bite needs to be determined. Clinical procedures such as partial glossectomy and orthodontic treatment are available, but guides to natural regression, criteria for intervention, and optimal age for intervention have yet to be established. (5 refs.) E. L. Rowan.

2444 SMITH, W. KING. Goldenhar syndrome (Oculo-auriculovertebral dysplasia). In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 213-214.

Marked ear deformities, an epibulbar dermoid, an upper lid coloboma, micrognathia, vertebral anomalies, and MR were characteristic of an 18-year-old girl with the Goldenhar syndrome. (No refs.) - E. L. Rowan.

2445 HALL, JUDITH G. Mandibulo-facial dysostosis (Treacher Collins-Franceschetti syndrome). In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 215-216.

Mandibulo-facial dysostosis in 2 patients was characterized by malar hypoplasia, antimongoloid slant of eyes, absent cilia and colobomata of lower lids, microtia, ear deformities, micrognathia, and mild to moderate MR. Both the mother and maternal grandfather of the first patient showed similar but less severe deformities while the second case probably represented a fresh mutation. (No refs.)

E. L. Rowan.

2446 HERRMANN, JURGEN, & OPITZ, JOHN M. An unusual form of acrocephalosyndactyly. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 39-42.

A previously unreported malformation syndrome is described in a boy with severe deformities of skull and limbs, short stature, and MR. He has a tall, bitemporally flattened head, hypertelorism, strabismus, micrognathia, ear anomalies, and a large bone defect in the posterior parietal area. Hands show soft tissue syndactyly, and there is only one digit on each foot. The father was 36 years old at the time of his wife's conception, and the patient's condition may represent a new mutation. (No refs.) - E. L. Rowan.

2447 AASE, JON M. Oto-palato-digital syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 43-44.

A woman and her 2 sons have identical clinical syndromes including characteristic "pugilistic" facies, small ear canals with conductive deafness, submucosal palatal clefts, and digital anomalies. Both boys are mildly MR. This oto-palato-digital syndrome seems determined by a dominant mode of inheritance rather than the recessive mode suggested in the past. (No refs.) - E. L. Rowan.

2448 GROSSE, FRANK REINER, HERRMANN, JURGEN, & OPITZ, JOHN M. The F-form of acropectoro-vertebral dysplasia: The F-syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 48-63.

Eight members of the "F" family showed a previously undescribed syndrome of acropectoro-vertebral dysplasia which consists of carpal and tarsal synostoses, deformity and syndactyly of the first and second fingers, postaxial toes, webbing between toes, prominent sternum, spina bifida occulta, minor craniofacial anomalies, and learning difficulties. The syndrome appears to be dominantly transmitted and results from both ectodermal and mesodermal defects beginning during the fourth post-ovulatory week. (12 refs.)

E. L. Rowan.

2449 DIEKER, HANS, & OPITZ, JOHN M. Associated acral and renal malformation. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 68-77.

Three unrelated patients manifest anomalies of limbs and kidneys which may be considered to be a formal genesis syndrome. All show syndactyly, brachydactyly, and ectrodactyly postulated to result from faulty segmentation of the hand or foot plate into digits. All 3 show unilateral renal agenesis, and in one there is a duplication of the contralateral kidney. Two show pectus excavatum, and two are MR. Advanced paternal age in 2 of the cases suggests a new mutation as the cause of the disorder. (21 refs.) - E. L. Rowan.

2450 HERRMANN, JURGEN, FEINGOLD, MURRAY, TUFFLI, GORDON A., & OPITZ, JOHN M. A familial dysmorphogenetic syndrome of limb deformities, characteristic facial appearance and associated anomalies: The "Pseudothalidomide" or SC-syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 81-89.

Pairs of siblings from 2 different families have a similar dysmorphogenetic syndrome of limb deformities, characteristic facies, and

MR. Clinically, these patients show symmetrical tetraphocomelia, flexion contractures, facial anomalies (capillary hemangiomata, hypoplastic cartilage of ears and nose, micrognathia, scanty silver-blond hair, and cloudy corneas), intrauterine growth retardation, and probable MR. The family pattern suggests an autosomal recessive mode of inheritance. (24 refs.) - E. L. Rowan.

- 2451 TENTAMY, SAMIA, & HALL, JUDITH G. Carpenter syndrome (Acrocephalopolysyndactyly). In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part III. Limb Malformations.* (Birth Defects Original Article Series, Volume 5, Number 3.) New York, New York, 1969, p. 204-206.

The patient in whom Carpenter's syndrome had first been described was reevaluated at age 7. No changes (except surgical corrections) were noted in the basic picture of acrocephaly, brachysyndactyly of the fingers, polysyndactyly of the toes, hypogenitalism, obesity, and MR. This girl also had pseudopapilledema, generalized aminoaciduria, and probable patent ductus arteriosus. (1 ref.) E. L. Rowan.

- 2452 HERRMANN, JURGEN, & OPITZ, JOHN M. Dermatoglyphic studies in a Rubinstein-Taybi patient, her unaffected dizygous twin sister and other relatives. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 22-24.

Finger anomalies and especially dermatoglyphic patterns were compared in a 12-year-old girl with Rubinstein-Taybi syndrome and her unaffected dizygous twin sister. The patient had a very low total ridge count (1/12 normal female value), but this trend was also noted in the twin (1/2 normal) and in an older sister (1/4 normal). The proband also had bilateral proximal axial triradii (also on father's left hand) and bilateral fourth interdigital distal loops. Double dermatoglyphic patterns were not found. The family distribution of total ridge count values suggests a multifactorial genetic etiology of this syndrome. (2 refs.) - E. L. Rowan.

- 2453 FILIPPI, GIORGIO. Rubinstein-Taybi syndrome in a Negro. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 208-210.

A Negro boy was diagnosed as having the Rubinstein-Taybi syndrome on the basis of profound MR, delayed physical development, and broad thumbs and great toes. Facies were characteristic and there was a sternal deformity. X-ray examination showed the distal phalanges to be tilted and bone age to be retarded. (No refs.) - E. L. Rowan.

- 2454 FILIPPI, GIORGIO. Rubinstein syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 211-212.

Rubinstein-Taybi syndrome was present in an 11-year-old boy with MR, short stature, hirsutism, syndactyly, and broad thumbs and toes. X-rays showed marked retardation of bone maturation, and the phalanges were strikingly broad and short. (No refs.) - E. L. Rowan.

- 2455 ZELLWEGER, HANS. The HHHO or Prader-Willi syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 15-17.

Over 100 cases of the HHHO (hypotonia, hypomenia, hypogonadism, obesity) or Prader-Willi syndrome have appeared in the literature. Two phases of the disease are noted. Newborns and young infants show amyotonia, poikilothermia, areflexia, nonreactivity, and hypogonadism. After weeks or months, these children become responsive and alert, but gradually pass on to a second phase characterized by hyperphagia and obesity, MR, delayed motor and speech development, emotional lability, small stature, and delayed bone maturation. EEG, karyotype, and muscle biopsy examinations have been nonspecific, and the condition is probably not hereditary. The etiology is unknown, but a hypothalamic defect is plausible. (6 refs.) - E. L. Rowan.

2456 MURDOCH, J. LAMONT. Prader-Willi syndrome. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes.* (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 213.

A 12-year-old Amish girl presented with obesity, short stature, small hands and feet, and strabismus. Motor and mental development were slow. Biochemical investigation disclosed a diabetic glucose-tolerance test and hyperlipidemia. A diagnosis of the Prader-Willi syndrome was made. (No refs.) - E. L. Rowan.

2457 HANISSIAN, ARAM S., & *SUMMITT, ROBERT L. Smith-Lemli-Opitz syndrome in a Negro child. *Journal of Pediatrics*, 74(2): 303-305, 1969.

A Negro male infant exhibited most of the malformations found in the Smith-Lemli-Opitz syndrome plus tibial torsion with metatarsus varus, paralysis of the palate and face, an aberrant right subclavian artery, premature fusion of the ossification centers of the sternum, bilateral vesicoureteral reflux, and a hypoplastic right kidney. Additional findings include a 15-degree rotation of the ears, symmetrical head, sparse hair, hypoplastic scrotum, undescended testes, and a bridged simian crease on the left palm. Digital patterns showed a simple arch on the first and second fingers of the right hand and first finger of the left hand, a whorl on the fourth finger of the right hand, and an ulnar loop on all other fingers. The S did not vomit, had no syndactyly, possessed a normal karyotype, and had a normal plasma growth hormone response to infusion of l-arginine monochloride. The family history was non-contributory. This is the first reported case in a Negro. (7 refs.) - A. Huffer.

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2458 BERGADA, C., FARIAS, N. ELVIRA, ROMERO DE BEHAR, BEATRIZ M., & CULLEN, M. Abnormal sex chromatin pattern in cryptorchidism, girls with short stature and other endocrine patients. *Helvetica Paediatrica Acta*, 24(4):372-377, 1969.

The incidences of abnormal sex chromatin in boys with cryptorchidism, girls with short stature, and other patients with endocrine problems admitted to the Children's Hospital of Buenos Aires during a 4-year period were

1/88 for cryptorchidism, 1/9 short girls, and 1/125 for all boys and 1/68 for all girls. The incidence rates found in this selected group of patients were higher than those found in newborns. In newborns, 1/3,000 and 1/500, respectively, were the incidences for chromatin-negative girls and chromatin-positive boys. (31 refs.) - L. S. Ho.

Departamento de Endocrinología
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Buenos Aires, Argentina

2459 WINTER, JEREMY S. D., KOHN, GERTRUDE, MELLMAN, WILLIAM J., & WAGNER, SEYMOUR. A familial syndrome of renal, genital, and middle ear anomalies. *Journal of Pediatrics*, 72(1):88-93, 1968.

Four girls in one family have various types of renal dysgenesis associated with internal genital malformations and middle-ear anomalies. These familial data illustrate that congenital malformations of the female internal genitalia may frequently be associated with upper urinary tract abnormalities and may also include middle-ear deformities. The association of external ear deformities with anomalies of the urogenital tract has already been noted; however a familial syndrome of middle-ear deformities with renal and genital malformations has not been reported before. One of the 4 girls, only 2 of whom survived, had trisomy of the X chromosome and was EMR. An autosomal recessive gene is suggested as responsible for this familial occurrence, although environmental or maternal teratogenic factors could not be excluded. (18 refs.) - B. Bradley.

Children's Hospital of Winnipeg
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2460 COHN, JORGEN, & BAY-NIELSEN, ERIK. Hereditary defect of the sacrum and coccyx with anterior sacral meningocele. *Acta Paediatrica Scandinavica*, 58(3):268-274, 1969.

Congenital absence of the sacrum and the coccyx with symptoms of constipation, incontinence of urine, infection of the urinary tract and anal stenosis were found in 6 members of one family. Four meningoceles were best shown by air- or panto-paquemyelography. This disorder is transmitted as a female sex-linked dominant gene. (34 refs.) - L. S. Ho.

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- 2461 FRASER, G. R., FRIEDMANN, A. I., MARO-TEAUX, P., GLEN-BOTT, A. M., & MITTWOCH, U. Dysplasia spondyloepiphysaria congenita and related generalized skeletal dysplasias among children with severe visual handicaps. *Archives of Disease in Childhood*, 44(236):490-498, 1969.

Among 776 children with severe visual handicaps, 7 cases of unusual generalized skeletal dystrophies affecting primarily the vertebrate and epiphyses of the long bones were found. In several cases, the condition was identical to dysplasia spondyloepiphysaria congenita. High myopia often preceded retinal detachment and blindness. Primary cataracts and buphthalmos were also found. In 3 cases, MR was suspected. The syndrome is probably transmitted as an autosomal dominant trait; however, the question of whether all the cases represent a single genetic defect with different phenotypic variations or whether 2 or more distinct conditions are involved needs further study. (32 refs.)

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- 2462 KIRKHAM, T. H. Cervico-oculo-acusticus syndrome with pseudopapilloedema. *Archives of Disease in Childhood*, 44(236):504-508, 1969.

The cervico-oculo-acusticus syndrome was found in 2 of 126 patients with Duane's syndrome and pseudopapilloedema was found to be associated with this syndrome for the first time. The appearance of pseudopapilloedema remained unchanged over a prolonged period and no other complications such as neurological disorders, hemorrhages, or exudate occurred. The calibre of the retinal vessel was normal. Pseudopapilloedema is probably genetically associated with the cervico-oculo-acusticus syndrome. (20 refs.)

L. S. Ho.

The Royal Infirmary
Sheffield, England

- 2463 GARCES, LUDIVINA Y., BLANK, EUGENE, *DRASH, ALLAN L., & KENNY, FREDERIC M. Peripheral dysostosis: Investigation of metabolic and endocrine functions. *Journal of Pediatrics*, 74(5):730-737, 1969.

Two unrelated girls (CAs 4 11/12 yrs and 3 5/12 yrs) with peripheral dysostosis, advanced bone age, but normal height were

found to have no metabolic nor endocrine abnormalities. Evaluation of pituitary adrenocorticotrophic hormone (ACTH); thyroid-stimulating hormone; serum and urinary gonadotrophins; and urinary estrogens indicated that the basal 24-hour urinary 17-hydroxycorticosteroids (17-OHCS) and 17-ketosteroids were normal. The total urinary estrogens were normal in one child and moderately elevated in the other. There was a normal response of growth hormone to arginine infusion, insulin-induced hypoglycemia, and oral glucose. The impaired glucose tolerance of one child with inadequate insulin response to glucose load, and diabetes mellitus in the family history suggested that the patient had genetic prediabetes. The normal serum and urinary calcium and phosphorus levels suggested that the parathyroid function was normal in both. (26 refs.) - F. J. McNulty.

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- 2464 WESTLUND, KNUT. Mortality from congenital malformations of the central nervous system in Norway, 1951-65. *British Journal of Preventive and Social Medicine*, 23(1):28-33, 1969.

The mortality data compiled in Norway from 1951-65 on congenital malformations of the nervous system indicated that severe abnormalities were excessive in the northern counties. There were no significant urban-rural rate differences. Abnormality and spina bifida showed a female excess, while males had hydrocephalus in excess. The mortality rate/10,000 live-births decreased from the preceding 5-year period, and there was no association of mortality by month of birth. Congenital malformations of the nervous system were classified according to 4 groups: anencephalus; spina bifida and meningocele; congenital hydrocephalus; and other malformations of the nervous system or sense organs, such as microcephalus. In the mortality by sex, there was a female excess in the first and second groups as well as for all groups combined. The distribution of congenital malformations suggests an association with unfavorable socioeconomic conditions. In the mortality time-trend, the spina bifida rate for the 15-year period decreased, but the spina bifida death at age 1-3 years increased during the period 1961-65 when compared to 1956-60. Hydrocephalus in the mortality time-trend had a rate increase during the 15-year period. (8 refs.) - F. J. McNulty.

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2465 JAMES, WILLIAM H. Central nervous system malformation stillbirths, maternal age and birth order. *Annals of Human Genetics*, 32(3):223-236, 1969.

The frequency of all stillbirths and of stillbirths due to anencephaly, spina bifida, and hydrocephalus in England, Scotland, and Wales during the period 1949-1964 was analyzed statistically with respect to maternal age and birth order. The proportion of CNS stillbirths diminishes with both maternal age and parity. Perinatal mortality data indicate that this decline in hydrocephalic stillbirths is due either to a fertility differential such that hydrocephalic-stillbirth-prone sibships are less fertile than other stillbirth-prone sibships, or to the existence of negative effects of birth order or maternal age. In anencephalic stillbirths there are probably negative birth order and negative maternal age effects operating independently and the same is possibly true in spina bifida stillbirths. (40 refs.)

M. G. Conant.

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2466 JAMES, WILLIAM H. Stillbirth, neonatal death and birth interval. *Annals of Human Genetics*, 32(2):163-172, 1968.

Although a study of 98% of all live-births from March 3-9, 1958 and 94% of all perinatal deaths for March, April, and May 1958 in England, Scotland, and Wales showed some relation between stillbirth, neonatal death, and birth interval, it was concluded that when a large sample is pooled, both stillbirths and neonatal deaths are associated with both long and short birth intervals; within sibships, brief birth intervals are not associated with stillbirth; stillbirths are associated with long birth intervals within sibships, although the cause for this is uncertain; and the relation of birth interval and neonatal death is unclear. In studying the relation among stillbirth, neonatal death, and birth interval, the data collected in the Perinatal Mortality Survey of 1963 were used: a total of 16,377 live-births and 7,117 deaths. Selection criteria were at least 2 prior pregnancies; no previous abortion, ectopic pregnancy, stillbirth, or neonatal death; parity, result of present confinement; and rank order within the sibship of length of birth interval. Statistical studies of relations were made in terms of live-births, stillbirths, neonatal deaths, live-births plus stillbirths, and live-births plus neonatal deaths. The ratio of stillbirth to live-birth and of

neonatal death to live-birth was determined. A trend which related stillbirth to long birth interval appeared, and it was postulated that factors of maternal disease, including diabetes, hypertension, thyroid disorder, syphilis, or others might contribute. Short birth intervals appeared, in one survey, to have some relation to neonatal death, but was probably a statistical artifact. Further study of birth interval and neonatal death rate is indicated. (27 refs.) - C. A. Rizvi.

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2467 Anencephalus and spina bifida. *British Medical Journal*, 2(5649):67-68, 1969. (Editorial)

Large population studies of spina bifida and anencephalus have shown a familial incidence of these malformations and significant regional variations; however, in no case have both members of a pair of twins been affected. A polygenic mode of inheritance is possible and studies of extended families might clarify this point; nevertheless, more attention should be paid to environmental influences. (11 refs.) - E. L. Rowan.

2468 HOROWITZ, I., & *McDONALD, A. D. Anencephaly and spina bifida in the Province of Quebec. *Canadian Medical Association Journal*, 100(6):748-755, 1969.

Anencephaly and spina bifida occurring in live-births and stillbirths in the Province of Quebec between 1956 and 1965 were ascertained as fully as possible and studied in relation to ethnic, geographic, seasonal, and certain maternal factors. The 2 neural tube defects were found to behave similarly in respect to these factors, suggesting a common etiology. For the period 1961-65, during which ascertainment was considered satisfactory, the rate of anencephaly was 1.44/1,000 live-births and that of spina bifida 1.87. These rates were lower than those observed in Britain but higher than those reported from France. Neural tube defects as a whole occurred as commonly in families of French as in those of British origin living in Quebec (3.4 and 3.2, respectively). Much lower rates were found among Jewish families (0.7 according to ethnic origin and 0.9 according to religion). No regional differences were detected, but rates were generally higher in urban than in rural areas. Rates in socio-economically favored districts were lower than in remaining areas. There appeared to

be minor seasonal fluctuations that were not of a statistically significant order. Rates in first pregnancies were higher than in subsequent pregnancies. All 41 twins with anencephaly were discordant for a neural tube defect. Of 20 twin pairs with spina bifida, 2 were concordant, of which one only could have been monozygotic. Since there was no indication of early fetal loss of monozygotic twins the low rate of concordance found in this and previous studies suggests that genetic factors are unimportant etiologically. There are few clues as to the nature of any possible environmental cause, but a latent or chronic maternal infection acquired before maternity, for which there is no direct evidence, offers a plausible hypothesis. (50 refs.) - *Journal summary*.

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2469 CHRISTAKOS, ARTHUR C., & SIMPSON, JOE LEIGH. Anencephaly in three siblings. *Obstetrics and Gynecology*, 33(2):267-270, 1969.

Three anencephalic stillborn infants and 2 spontaneous abortions occurred in a family of Anglo-Saxon descent. The first, third, and fifth pregnancy terminated with spontaneous delivery of an anencephalic child. Although Bendectin had been given during 2 of these pregnancies and during one of the abortions, it is felt that drugs did not play a prominent role. Neither viral infections nor other environmental factors were detected that might explain the recurrent anencephaly. Analysis of the family pedigree in which the previous generation on both sides had MR suggests the possibility that anencephaly has an autosomal recessive inheritance basis--a single dose of a low penetrance gene may cause MR while a double dose may cause anencephaly. (31 refs.) - *A. Huffer*.

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2470 NASH, D. F. ELLISON. Urinary problems of spina bifida. *Developmental Medicine and Child Neurology*, 11(1):106-108, 1969. (Annotation)

Infection and incontinence are the major urinary problems of spina bifida, and resultant

pyelonephritis and hypertension are the major causes of death. Frequent micturition with a high rate of flow has reduced the stasis which often led to infection, and bacterial identification and the subsequent use of specific antibiotics have reduced morbidity. Ileal loop diversion relieves incontinence but should not be attempted until it is established that the child cannot otherwise obtain control. (16 refs.) - *E. L. Rowan*.

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2471 Special needs of children handicapped by spina bifida. *Nursing Mirror*, 128 (23):13, 1969.

Children with spina bifida require special educational facilities because of their physical handicaps. Effective bladder and bowel control must be established, and a nurse or pediatrician must be alert for infection, pressure sores, or signs of a blocked ventriculoatrial shunt. Both physical problems and emotional difficulties associated with separation, incontinence, or paraplegia must be dealt with so that the child may develop his only real asset--his intellect. (No refs.) - *E. L. Rowan*.

2472 NEW YORK UNIVERSITY SPINA BIFIDA STUDY GROUP. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, 147 p.

To reduce the morbidity and mortality of children with spina bifida manifesta, rehabilitation programs should be interdisciplinary and should begin at birth. Neonates and young infants require pediatric, neurological, and neurosurgical services, and their parents require counseling. The papers presented in this monograph are the final report of the New York University Spina Bifida Study Group to the Vocational Rehabilitation Administration. This study group is an interdisciplinary group of medical and ancillary specialists. Spina bifida is defined, and data on incidence rates and ecological factors are presented. Topics covered include statistics on clinic services to spina bifida patients, pediatric and neurological management, incontinence, orthopedic care, muscle activity, intellectual functioning, verbal behavior, rehabilitation, family counseling and education, and recreation. This monograph should be of interest to pediatricians,

urologists, neurosurgeons, orthopedic surgeons, psychiatrists, ophthalmologists, psychologists, social workers, nurses, physical therapists, occupational therapists, speech and hearing therapists, and educators. (165 refs.) - J. K. Wyatt.

CONTENTS: Definition, Nomenclature and Incidence of Spina Bifida with Special Reference to Ecological Factors (Swinyard & Shahani); Spina Bifida Clinic-Patient Population (Greenspan & Poole, Jr.); Pediatric Care of the Child with Spina Bifida Manifesta (O'Hare & Greenspan); The Neurologic and Neurosurgical Management of Spina Bifida Manifesta (Berman, Mathews, & Ransohoff); The Relationship of Non-Progressive Hydrocephalus to Intellectual Functioning in Children with Spina Bifida Cystica (Badell-Ribera, Shulanan, & Paddock); Urologic Problems in Children with Myelomeningocele (Morales); Orthopedic Care of the Child with Spina Bifida (Tzimas); An Electromyographic Study of Children with Spina Bifida Manifesta (Chantraine, Lloyd, & Swinyard); The Sphincter Ani Externus in Spina Bifida and Myelomeningocele (Chantraine, Lloyd, & Swinyard); Spina Bifida with Myelomeningocele: Evaluation of Rehabilitation Potential (Badell-Ribera, Swinyard, Greenspan, & Deaver); Verbal Behavior in Children with Spina Bifida (Diller, Paddock, Badell-Ribera, & Swinyard); A Comparison of Intellectual Development of Children with Spina Bifida with their Normal Siblings (Paddock); The Management of Incontinence in Children with Spina Bifida and Myelomeningocele (Mihalov, Morales, Badell-Ribera, & Swinyard); The Importance of Family Education and Counseling (Swinyard); A Therapeutic Recreational Program for Children with Spina Bifida and Myelomeningocele (Carr, Mihalov, & Swinyard); Vocational Rehabilitation Case Load and Potential of Patients with Spina Bifida and Myelomeningocele (Badell-Ribera, Siegal, & Swinyard).

- 2473 SWINYARD, CHESTER A., & SHAHANI, BHAGWAN T. Definition, nomenclature and incidence of spina bifida with special reference to ecological factors. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 3-9.

Although the etiological factors which cause human spina bifida have not been identified, the causative factors appear to be profoundly influenced by an individual's genetic constitution as well as by ecological factors. The most frequent type of spina bifida is spina

bifida manifesta or myelomeningocele, a condition in which the defective spinal cord and the meninges are visible on the back with the skin attached to the side of the protruding mass. Children with this malformation usually have muscle weakness, skin insensitivity below the weakness, defective bowel and bladder innervation, and varying degrees of ventricular dilation with or without hydrocephalus. The incidence of spina bifida is approximately 2-3/1,000 live-births, and it is more frequent and tends to be more severe in females than in males. Incidence is higher in whites than in non-whites, in first pregnancies, and in plural births. It appears to be predisposed by urbanity, and there is some evidence of higher incidence in industrial areas and among lower socioeconomic classes. The incidence is higher in births between December and May and in siblings of infants with central nervous system malformations. (33 refs.) - J. K. Wyatt.

- 2474 GREENSPAN, LEON, & POOLE, CHRISTIAN H., JR. Spina bifida clinic-patient population. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 10-13.

One hundred and sixty-five patients (CA over 15 mos; 123 in-patients, 42 out-patients) received rehabilitation services in a spina bifida clinic over a period of 3 years and 9 months. Each patient was completely evaluated to assess the multiple defects which stem directly from the malformation as well as behavioral problems and psychosocial adjustment. In-patient evaluation was recommended because it allowed for closer observation of the child and of parent-child relationships. Each child averaged 2.6 admissions to the clinic which lasted an average of 34.1 days. Approximately 200 ventriculojugular shunts or shunt revisions were required, and ureteroileostomy was performed in 14 patients. Braces were provided for 78 children, 25 of whom also received appropriate wheelchairs. The 42 patients treated exclusively on an out-patient basis had an average of 5.2 visits, and the total group of patients had an average of 7.2 out-patient visits. Periodic out-patient re-examinations were used to evaluate the effectiveness of home programs, detect changes related to growth and development, continue parent counseling, and recommend additional in-patient therapy. (No refs.) - J. K. Wyatt.

2475 O'HARE, DONNA, & GREENSPAN, LEON. Pediatric care of the child with spina bifida manifesta. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 14-16.

The pediatrician's role in the rehabilitation of children with spina bifida manifesta is to provide a total individualized medical program which may include the mobilization of specialized services, cooperation with medical specialists, and parental and family counseling. The primary concern during the neonatal period is the removal of the sac and/or correction of hydrocephalus by neurosurgery. During the first year, the child must be observed to detect evidence of increased intracranial pressure or infection and, when the child has had a shunting procedure, to identify a malfunctioning or contaminated shunt. The child should receive all recommended immunizations, and parents should be taught to provide meticulous skin care. During childhood, specialized care by an orthopedist and physiatrist is needed to prevent or reduce deformities related to imbalance of muscle groups in the lower extremities and to make the child as independent as possible. The physical limitations and hygiene state of the school-age child with spina bifida manifesta as well as architectural barriers, transportation barriers, and negative school personnel attitudes may limit acceptance into educational programs. The pediatrician should cooperate with other specialists to assure school admission and appropriate educational experiences. Broad rehabilitation objectives for children with spina bifida manifesta include maximal independence, appropriate educational experiences, and opportunities to pursue a vocation related to their interests and abilities. (No refs.) - J. K. Wyatt.

2476 BERMAN, PETER H., MATHEWS, ERNEST S., & RANSOHOFF, JOSEPH. The neurologic and neurosurgical management of spina bifida manifesta. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 17-21.

To provide optimal neurological and neurosurgical care for infants with spina bifida, a multidisciplinary diagnostic evaluation which recognizes the interrelations of their multiple problems is required. During the

neonatal and early infancy periods, hydrocephalus and meningitis must be detected, evaluated, and treated. When the meningitis sac is intact, the surgical closure of all uncomplicated spinal defects should be delayed until coincidental complications of hydrocephalus and/or meningitis have been alleviated. However, in cases where there is leakage of cerebrospinal fluid from the meningeal sac, the closure of spinal defects should be given immediate priority. Diagnosis of meningitis in the newborn usually requires examination of the spinal fluid, and treatment is by parenteral antibiotic therapy for a minimum of 10 days. Echoencephalography and air ventriculography are used during initial evaluation for hydrocephalus. Management is by ventriculoatrial shunt (in the absence of infection) or by appropriate antibiotic therapy and necessary ventricular drainage (in the presence of infection). (8 refs.) - J. K. Wyatt.

2477 MORALES, PABLO A. Urologic problems in children with myelomeningocele. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 32-44.

Children who have had primary surgery on the myelomeningocele and stabilization of ventricular dilation and/or hydrocephalus experience urologic problems resulting from lower motor neuron impairment of bladder and urethra. Children with "neurogenic" bladders have some patterns of urinary incontinence and a lack of sensation of bladder fullness or urethral urinary flow. Impaired bladder innervation may also result in excessive residual urine after manual efforts to empty the bladder, bacteriuria, trabeculation, and unilateral or bilateral vesico-uretic reflux. Treatment should include a urologic evaluation consisting of an intravenous pyelogram, a cystourethrogram, a urine culture, sensitivity tests, and measurements of residual urine. Evaluations should be made during the first year and annually thereafter. Renal damage can be prevented if preventive measures eliminate residual urine, control urinary infection, and, when required, effect permanent urinary diversion. Incontinence management includes periodic scheduled voidings, and the use of urinary bags, rubber pants, and absorbent cellulose. (6 refs.) J. K. Wyatt.

2478 CHANTRAINE, ALEX, LLOYD, KATHLEEN, & SWINYARD, CHESTER A. An electromyographic study of children with spina bifida manifesta. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 66-78.

Of 53 children (32 males, 21 females; CA 2 to 18 yrs) born with myelomeningocele, 12 had good to average lower limb muscle power (Group I), 6 had average to fair lower limb muscle power (Group II), 14 had good muscle power above the knee and zero below the knee (Group III), and 21 had poor to zero lower limb muscle power (Group IV). Electromyographic studies revealed that although increased insertion potentials were found in all groups, they were more common in Groups II and III. Fasciculations were found in 45% of the Ss and occurred in all groups. Muscles which tested fair evidenced the greatest frequency of fasciculation potentials. Fibrillation potentials occurred in 41% of the Ss in Groups II, III, and IV, but were not found in Ss in Group I. They frequently occurred in muscles with minimal fibrosis which tested from zero to poor. (24 refs.) - J. K. Wyatt.

2479 CHANTRAINE, ALEX, LLOYD, KATHLEEN, & SWINYARD, CHESTER A. The sphincter ani externus in spina bifida and myelomeningocele. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 79-87.

An analysis of the sphincter ani externus activity of 50 incontinent patients (CA 2 to 22 yrs; 30 males, 20 females) with spina bifida who had neurosurgical care for the myelomeningocele confirms the hypothesis that in a high or short spina bifida lesion, the conus medullaris may be intact and exert regulatory control. Methods of evaluation included digital examination, bulbocavernosus reflex, anal reflex, quantitative assessment of sphincter tension, electromyographic recordings, and urologic and neurological examinations. Ss who appeared to have similar

lesions had different electromyographic responses, and Ss with a positive bulbocavernosus reflex had electromyographic findings which showed a complete inference pattern with a 300 to 400 microvolt amplitude. Electromyographic findings in the sphincter ani and sphincter urogenitalis suggest that the functional status of the sphincter ani parallels that of the sphincter urogenitalis. When electromyographic potentials indicated loss of motor units in the muscle, there was a significantly below normal amount of quantitative tension. (18 refs.) - J. K. Wyatt.

2480 MEHREGAN, AMIR H. Elastosis perforans serpiginosa: A review of the literature and report of 11 cases. *Archives of Dermatology*, 97(4):381-393, 1968.

Clinical features and histopathologic changes of elastosis perforans serpiginosa (EPS) are reviewed from observations on 90 cases recorded in the literature and 11 additional cases. Among these 101 cases, 90% of the patients are under 30 years of age; 78 Ss are men; and EPS is found scattered all over the world. Eruption is, in most cases, confined to single anatomical regions such as nape and sides of the neck, upper extremities, face, lower extremities, and trunk. The primary lesion is a slightly erythematous or skin-colored keratotic papule (2-5 mm) with a small central area of scaling. The clinical course of the disease appears unpredictable. New lesions continue to develop as the old ones disappear. Treatments with dry ice and electrosurgery are unsatisfactory. Stripping the surface keratinous material with cellophane tape was effective in one of the 11 cases. Biopsies reveal areas of perforations in the form of narrow canals, and these areas are in completely transepidermal, perifollicular, or transfollicular positions. This and other data indicated that EPS may be another example of the transepidermal elimination phenomenon which is found in some cutaneous disorders. In 26% of the cases, the disease is associated with mongolism, the Ehlers-Danlos syndrome, osteogenesis imperfecta, pseudoxanthoma elasticum, or Marfan's syndrome. (74 refs.) - L. S. Ho.

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Gross brain disease (postnatal)

2481 AMMANN, A. J., CAIN, W. A., ISHIZAKA, K., HONG, R., & GOOD, R. A. Immunoglobulin E deficiency in ataxia-telangiectasia. *New England Journal of Medicine*, 281 (9):469-472, 1969.

An IgE deficiency was demonstrated in 11 of 16 patients with ataxia-telangiectasia. No relation to age or to the level of IgG, IgM, or IgD was found. Nine out of 11 patients with IgE and IgA deficiency had recurrent sinopulmonary infections of varying degrees. The varying times of onset of recurrent sinopulmonary infection may be a function of immunologic attrition. There may be a delay between the loss of IgE or IgA (or both) and the manifestations of this deficiency. Particular vulnerability to sinopulmonary disease in patients with ataxia-telangiectasia attends the lack of both IgA and IgE. (30 refs.) - *Journal abstract*.

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2482 HULTBERG, BJORN, OCKERMAN, PER-ARNE, & ERIKSSON, ORJAN. Urinary amino acids in storage disorder: Mucopolysaccharidosis, Gaucher's disease and metachromatic leukodystrophy. *Metabolism*, 18(8):713-719, 1969.

Amino acid metabolism was shown to be disturbed in Hurler's disease and a pathological pattern of amino acid excretion exists in Hunter's disease, Gaucher's disease, and metachromatic leukodystrophy (ML). Urine specimens were examined for amino acids in 15 patients and 23 control children by a Technicon amino acid analyzer. There were 6 patients (CA 2-8 yrs; 5 girls and 1 boy) with mucopolysaccharidosis, type I (Hurler's syndrome); 1 male patient (CA 19 yrs) with type II, and 4 male patients (CA 3-8 yrs) with type I or II. Two male patients (CA 13-15 yrs) were heparin sulfate excretors with mental regression and mucopolysaccharidosis type III. Two female patients (CA 11 and 27 yrs) had type IV. One girl (CA 3 yrs) and 3 boys (CA 2, 15, and 16 yrs) had Gaucher's disease. Three patients, a girl (CA 4 yrs)

and 2 boys (CA 3 and 8 yrs) had MLD. There was an increase in serine, threonine, and tryptophan in mucopolysaccharidosis types I and II. Patients with types III and IV had normal values. Only 1 out of the 15 patients had generalized aminoaciduria. The excretion of serine and threonine was increased in patients with Gaucher's disease and those with MLD. A mechanism is proposed in which the covalent linkage between xylose in the mucopolysaccharide chain and serine in the polypeptide chain is split by a β -xylosidase. (17 refs.) - F. J. McNulty.

Department of Clinical Chemistry
University Hospital
Lund, Sweden

2483 GREENE, HARRY L., HUG, GEORGE, & SCHUBERT, WILLIAM K. Metachromatic leukodystrophy: Treatment with arylsulfatase-A. *Archives of Neurology*, 20(2):147-153, 1969.

A patient with late infantile metachromatic leukodystrophy was treated with intravenous and intrathecal infusions of arylsulfatase-A. Although the enzyme levels became normal in the liver, the preparation did not enter the brain and was not of clinical benefit. The patient showed no intolerance to the preparation when injected intravenously, but when injected intrathecally, he developed a temperature elevation within 4 hours which persisted for 8 hours. His neurologic state did not change during the infusion period nor during the 3 months following the study. The first infusion raised the serum levels of arylsulfatase-A in 10 minutes, after which the levels dropped to somewhat lower values. Eight hours after the injection, biopsy specimens of brain and liver were analyzed and showed no enzyme activity. No detectable light or electron microscopic changes occurred in the brain nor the liver with the enzyme treatment. (16 refs.) - F. J. McNulty.

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2484 FAHMY, ALY, CARTER, THERESA, PAULSON, GEORGE, & NANCE, WALTER E. A "new" form of hereditary cerebral sclerosis: An autosomal recessive neurologic disease with distinctive electron microscopic findings. *Archives of Neurology*, 20(5):468-478, 1969.

A brother and sister are reported who are affected with a slowly progressive neurologic disease which first became apparent in early childhood. Both show ataxia, speech difficulties, nystagmus, visual impairment, involuntary head movements, spasticity, and retinal defects. Examination of sural nerve biopsies revealed the inclusion of rod-shaped structures and zebra bodies in Schwann cells and the presence of concentric lamellar inclusions in macrophages. Nine siblings of the patients and their 56 cousins, nieces, and nephews are unaffected; however the parents are first cousins which suggests a mode of autosomal recessive transmission. The disease resembles Pelizaeus-Merzbacher syndrome except for the retinal changes, electron microscopic findings, and mode of inheritance. The unique morphological changes in the sural nerves as well as nonspecific abnormalities of nerve degeneration which are observed have been described as characteristics of nerve injury or of spontaneous or experimental degenerative or demyelinating diseases. (59 refs.) - M. G. Conant.

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Nashville, Tennessee 37203

2485 WATANABE, ITARU, McCAMAN, RICHARD, DYKEN, PAUL, & ZEMAN, WOLFGANG. Absence of cerebral myelin sheaths in a case of presumed Pelizaeus-Merzbacher disease. *Journal of Neuropathology and Experimental Neurology*, 28(2):243-256, 1969.

A 6-month-old infant with clinical symptoms of Pelizaeus-Merzbacher disease was shown, microscopically, to have no compact myelin sheaths in the biopsy samples of cortex and subcortical white matter. Some astrocytes containing peculiar crystalline dense bodies and a decreased number of oligodendrocytes were found in the white matter. The absence of compact myelin sheaths indicated a lack of proper myelination in this case rather than a demyelinating process. (16 refs.)

L. S. Ho.

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2486 O'BRIEN, JOHN. Generalized gangliosidosis. *Clinical Proceedings of Children's Hospital*, 25(2):40-52, 1969.

Gangliosidosis is a lipid and mucopolysaccharidosis storage disease in which the GM₁ ganglioside is present in more than one organ and a galactose-containing mucopolysaccharide (MPS) accumulates in the visceral organs. The clinical manifestations differ from Hurler's syndrome in that in generalized gangliosidosis, cataracts do not occur, and the clinical course is much more rapid. The humerus deformity and the periosteal cloaking are 2 of the best diagnostic radiological signs. The β -galactosidase activity was studied in patients, and the results demonstrated a tenfold deficiency of β -galactosidase in the brain. The activity of a number of other acid hydrolases were assayed, including acid phosphatase, β -glucosidase, and β -glucosaminidase. The activities of all these enzymes were increased in generalized gangliosidosis which indicates that the deficiency of β -galactosidase was specific, and other lysosomal enzymes were neither inhibited nor defective. Recent work has shown that a galactose-containing MPS which is structurally similar to keratosulfate accumulates in this disorder. Keratosulfate is a sulfate containing both galactose and N-acetylglucosamine. There is a similarity between the structure of the MPS and that of GM₁ because both contain galactose linked to hexosamine. The question arose as to whether β -galactosidase might cleave both compounds and whether its deficiency could lead to the accumulation of both. The keratosulfate was isolated from the liver of a patient with generalized gangliosidosis; cleavage of galactose from this compound by normal and affected livers was then tested. A tenfold lower rate of cleavage of galactose from the MPS in generalized gangliosidosis was found. (10 refs.)

F. J. McNulty.

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2487 SUZUKI, KUNIHICO, SUZUKI, KINUKO, & KAMOSHITA, SHIGEHICO. Chemical pathology of GM₁-gangliosidosis (generalized gangliosidosis). *Journal of Neuropathology and Experimental Neurology*, 28(1):25-73, 1969.

Chemical investigation and comparison of 5 brains and 3 livers and spleens from patients with cerebral GM₁-gangliosidosis with one brain and 3 livers and spleens from patients

of similar ages with Tay-Sachs disease (GM2-gangliosidosis) indicated that GM1-gangliosidosis can be classified as a distinct disease entity. The cerebral levels of the normal major monosialoganglioside (GM1) and its asialo-derivative were greatly increased in GM1-gangliosidosis, a situation analogous to that in Tay-Sachs disease and, in both diseases, the white matter showed signs of non-specific myelindegneration (probably a secondary effect). Membranous cytoplasmic bodies isolated from the brains of both diseases had very similar molecular compositions and livers and spleens of both gangliosidoses contained abnormally high concentrations of characteristic gangliosides. The visceral organs of GM1-gangliosidosis also contained excessive amounts of a mucopolysaccharide, possibly keratan sulfate, and a related sialomucopolysaccharide. The chemical data obtained in this study, using thin layer and column chromatography, indicate that cerebral GM1-gangliosidosis is a combined cerebral gangliosidosis and visceral mucopolysaccharidosis that may be related to the reported β galactosidase deficiency in this disease. (112 refs.) - M. G. Conant.

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- 2488 HUBAIN, P., ADAM, E., DEWELLE, A., DRUEZ, G., FARRIAUX, J. -P., & DUPONT, A. Etude d'une observation de gangliosidose a GM1 (Report of a case of GM1 gangliosidosis). *Helvetica Paediatrica Acta*, 24(4):337-351, 1969.

A female infant of consanguineous parents, first seen at the age of 6 months, showed psychomotor retardation, facial dysmorphism of the Hurlerian type, hepatosplenomegaly, vacuolated lymphocytes, and deformation of the first and second lumbar vertebrae. A diagnosis of generalized GM1 gangliosidosis was made. Death ensued at age 16.5 months, and electron microscopic examination of visceral cells revealed enlarged lysosomes containing a granular and fibrillar material. Increased amounts of GM1 ganglioside were found in the liver, spleen, kidneys, and brain, but β -galactosidase was absent. A review of 17 cases of this disease indicated that there are 2 forms of GM1 gangliosidosis: one which first appears at 0-5 months of age and involves hepatosplenomegaly, skeletal anomalies, and accumulation of GM1; the other which

appears at 7-14 months of age involves accumulation of GM3 and does not include hepatosplenomegaly or skeletal anomalies. (26 refs.) - M. G. Conant.

No address

- 2489 CUMINGS, J. N., THOMPSON, E. J., & GOODWIN, H. Sphingolipids and phospholipids in microsomes and myelin from normal and pathological brains. *Journal of Neurochemistry*, 15(2):243-248, 1968.

Human cerebral white matter and cortex from 5 normal brains, 2 cases of metachromatic leukodystrophy, 2 cases of Krabbe's disease, and 1 case of Tay-Sachs disease were examined. Myelin was isolated from the white matter and microsomes from the cortex using the 15% cesium chloride technique, and their sphingolipid and phospholipid contents were estimated by using thin-layer chromatography. Gangliosides were absent in normal myelin, but present in the microsomes and myelin of pathological preparations. In addition, the normal cerebroside:sulfatide ratio of 4:1 was found to be 1:20 in metachromatic leukodystrophy and 7:1 in Krabbe's disease. Preparations from Tay-Sachs disease brain showed a great increase in ganglioside (much of which is probably in the cytoplasmic membranous bodies) and an increase in trihexoside components. (19 refs.) - M. G. Conant.

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- 2490 SUZUKI, KINUKO, SUZUKI, KUNIHICO, & CHEN, GLORIA C. Morphological, histochemical and biochemical studies on a case of systemic late infantile lipidosis (Generalized gangliosidosis). *Journal of Neuropathology and Experimental Neurology*, 27(1):15-38, 1968.

Electron microscopic, histochemical, and biochemical studies were made on postmortem cerebral tissue (processed within 6 hrs after death) obtained from a 37-month-old male with systemic late infantile lipidosis. Membranous cytoplasmic bodies similar to those found in Tay-Sachs disease were abundant in the neuronal cytoplasm, and different membrane-bound vesicular cytoplasmic inclusions were found in glial cells. Macrophages found in the liver and spleen had tubular cytoplasmic inclusions unlike those found in the central nervous system. Oxidative enzymatic

activity was noted only on the periphery of the neuronal perikarya, dendrites and axons; however, acid phosphatase positive granules were abundant in the neuronal cytoplasm, and acid phosphatase activity was also seen in glial cells. Thin-layer chromatographic analysis of cerebral white and grey matter revealed an excessive accumulation of G₄ ganglioside and an abnormal ceramide hexoside pattern. In addition, the white matter showed moderate to severe demyelination, a finding which was supported chemically by the presence of low total lipids, low proteolipid protein, and large amounts of cholesterol ester. Degradative pathways of ganglioside metabolism are not yet established. (53 refs.) - M. G. Conant.

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- 2491 CARAKUSHANSKY, GERSON. The lipidoses. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 17, p. 956-978.

Mental disturbances are involved in the following hereditary lipid storage diseases: Niemann-Pick disease, metachromatic leukodystrophy, Tay-Sachs disease, juvenile amaurotic idiocy, generalized gangliosidosis, and Hurler's syndrome. Each disease seems to have a distinct defect of one of the lipid degradative enzymes in its etiology. Glucocerebrosidase-cleaving enzyme deficiency results in the accumulation of glucocerebrosidase in the reticuloendothelial system in Gaucher's disease, while the absence of sphingomyelinase probably causes an accumulation of sphingomyelin in the brain and visceral organs. Fabry's disease is characterized by patterned skin lesions, an onset at the time of puberty, and sex-linked dominance. Farber's lipogenesis usually occurs in early infancy and in most cases is lethal. Where genetic information is collected, autosomal recessive transmission is the most usual mode of inheritance. No specific treatment is available for any of the diseases. (164 refs.) L. S. Ho.

- 2492 EVANS, J. E., & McCLUER, R. H. The structure of brain dihexosylceramide in globoid cell leukodystrophy. *Journal of Neurochemistry*, 16(9):1393-1399, 1969.

Cerebral grey matter and basal ganglia from a formalin-fixed brain of a case of globoid

cell leukodystrophy were subjected to mild alkaline hydrolysis and sequential chromatography to isolate dihexosylceramide. The carbohydrate sequence in dihexosylceramide was shown to be galactosylglucosylceramide after partial degradation by mild acid hydrolysis and by methylation. Ozonolysis gave a free disaccharide moiety which was shown to be lactose by gas-liquid chromatography. The only long chain bases detected were 4-sphinganine and sphinganine, and the chain length of the fatty acids present ranged from 14 to 25 carbons. The structure of the dihexosylceramide isolated from this case of globoid cell leukodystrophy was established as lactosylceramide. It is suggested that the accumulation of this substance does not reflect any metabolic abnormality. (19 refs.)

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- 2493 HERMAN, MARY M., HUTTENLOCHER, PETER R., & BENSCH, KLAUS, G. Electron microscopic observations in infantile neuroaxonal dystrophy: Report of a cortical biopsy and review of the recent literature. *Archives of Neurology*, 20(1):19-34, 1969.

Electron microscopic examination of a frontal cortical biopsy from a 27-month-old female with the clinical symptoms suggestive of infantile neuroaxonal dystrophy is reported. Large axonal swellings filled with a sponge-like network of tubular structures usually composed of compactly arranged smooth membranes were observed. Sometimes these swellings participated in axodendritic or axo-axonic synaptic complexes, suggesting a possible relation between the smooth membranes and synaptic vesicles. The axonal swellings may represent either a sustained regenerative response of the axoplasm to some sort of injury or a neoformation of axoplasmic membranes unrelated to axonal injury. Most cortical structures, including neuronal cell bodies, dendrites, and most myelinated fibers, are preserved and the fact that the lesion is largely confined to the axon may account for the profound dementia associated with this disorder. (93 refs.) - M. G. Conant.

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- 2494 COLLINS, GEORGE H., COWDEN, RONALD R., & NEVIS, ARNOLD H. Myoclonus epilepsy with Lafora bodies: An ultrastructural and cytochemical study. *Archives of Pathology*, 86(3):239-254, 1968.

Cerebral biopsy material removed from a teenage boy with myoclonus epilepsy was studied by use of histological, cytochemical, and ultrastructural techniques. The material was found to include neurons containing Lafora bodies, neurons with other abnormalities involving the cytoplasm and nucleus, and glial cells with abnormal cell processes. The Lafora bodies have as a fundamental unit a fibril sometimes appearing as a tubular structure, and developing in association with endoplasmic reticulum and ribosomal material. The central core and the peripheral zone showed qualitative similarities and quantitative differences. The Lafora bodies showed strong staining reactions for carbohydrates and weak reactions for protein, a minimal amount of acid mucopolysaccharide, and a greater resistance to carbohydrate than to protein enzymatic digestion. The protein moiety is sensitive to chymotrypsin and pepsin. The principal material contained in the Lafora body may be glycoprotein. (54 refs.)
M. G. Conant.

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- 2495 SUZUKI, JIRO, & TAKAKU, AKIRA. Cerebrovascular "Moyamoya" disease. *Archives of Neurology*, 20(3):288-299, 1969.

Cerebrovascular "moyamoya" disease is a net-like collection of blood vessels over the base of the brain in association with occlusion of the internal carotid artery. It occurs commonly among the Japanese and primarily in young girls. In this series of 20

cases, the picture was variable as children most often presented with a history of transient ischemic episodes (repetitive hemiplegia) and MR and, on angiography, the lesion was seen to progress (the network increased as the carotid became more occluded). In adults the onset was associated with seizures and subarachnoid hemorrhage and there was no progression demonstrated on cerebral angiography. The picture is somewhat similar to that of pulseless (Takayasu's) disease and much work needs to be done to clarify the etiology. (10 refs.) - E. L. Rowan.

Department of Neurosurgery
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Sendai, Japan

- 2496 GALLI, CLAUDIO, & GALLI, DANIELA RE CECCONI. Cerebroside and sulfatide deficiency in the brain of "Jimpy mice," a mutant strain of mice exhibiting neurological symptoms. *Nature*, 220(5163):165-166, 1968.

The brains of 15 Jimpy mice were examined with quantitative microanalytical procedures to determine biochemical changes involving brain lipids and were found to have deficiencies of cerebroside and sulfatides. The cerebroside content in Jimpy mouse brain was less than 0.1% compared to 3.5% for control mice. The sulfatide content was less than 0.1% in Jimpy mouse brain compared to 1.7% for control brains. This suggests that a defect in the synthesis of these 2 glycolipids as well as impairment of myelin formation may be the basic alterations in the Jimpy mouse brain. (11 refs.) - F. J. McNulty.

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Psycho-environmental

2497 PATTON, ROBERT GRAY, & GARDNER, LYTT I. Short stature associated with maternal deprivation syndrome: Disordered family environment as cause of so-called idiopathic hypopituitarism. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 1, p. 77-89.

Growth retardation and disturbances in intellectual, motor, and emotional development are associated with a maternal deprivation syndrome which occurs in some children. These children often show stereotypic motor activity, especially an infantile posturing of the extremities, rumination, polydipsia, bulimia, and withdrawal. Improvement of environment (intensive care in the hospital, replacement of foundling home care by foster home care, or a change of maternal-infant relations) produces striking initial improvements in physical, mental, and intellectual development. Nevertheless, these children usually remain below age norms during the follow-up period. Malnutrition does not seem to be a primary factor of growth failure; on the contrary, mental depression, which has a profound effect on the gastrointestinal function of the infant, may be the major contributing factor to growth failure. Depression may also affect the functions of the anterior hypothalamus in such a way as to interfere with release and production of the anterior pituitary hormones. Many deprived children show abnormal responses to metyrapone and have inadequate increases in serum growth hormone after hypoglycemic stimulus. Thus, many cases of idiopathic hypopituitarism may, in reality, be instances of the maternal deprivation syndrome. Treatment for this disorder includes psychotherapy and supportive counseling of the mothers, good foster homes, or adoptive placement. (58 refs.) - L. S. Ho.

children were invited to make referrals of 4-year-olds eligible for the Early School Admissions Project. Children were admitted if their families could be characterized by 3 of 10 indices of cultural deprivation. Over the 3 years of the project, 842 children were examined. Public health nurses serving the 4 centers interviewed each parent using a structured standardized interview. Attending physicians using standardized neurologic examinations discovered 66 children with severe medical problems who were subsequently excluded from the project. Of the remaining children, 8 had definite major neurological defects, and 1.3% had definite neurological findings. Strabismus was found in 2.4%, and latent strabismus was revealed in 0.8%. Health histories showed that 3.8% of the children had fits or convulsions, 1.7% had "staring" or "fainting" spells, 57% still wet their beds at night, 14.5% stuttered or stammered, 25.3% sucked their thumbs, whirling was reported in 19.9% of the children, rocking was reported in 8.6% and head-banging was reported in 3%. Only 35% of these children were reported to be free of habits that indicated emotional tension. Other studies have documented the increased frequency of medical, social, and emotional problems in culturally disadvantaged children. The main purposes of the Early Admissions Project were to offer these children a program of activities that would prevent them from falling further behind their peers, to make known the medical complexities of low-income children entering the classroom, and to illustrate the effects of cultural deprivation in abnormal behavior and educational underachievement. (22 refs.) B. Parker.

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2498 STINE, OSCAR C., SARATSIOTIS, JOHN S., & FURNO, ORLANDO F. Selected neurologic and behavioral findings of children entering an early school admissions project from culturally deprived neighborhoods. *Journal of School Health*, 39(7):470-477, 1969.

Four schools which served neighborhoods with a high proportion of culturally deprived

2499 JACKSON, R. N. Urban distribution of educable mental handicap. *Journal of Mental Deficiency Research*, 12(4):312-316, 1968.

The incidence of MR was found to be higher in city areas characterized by overcrowding, poor infant care, endemic disease, high birth rate, and social disorganization. Special school referral rate was highest in areas where: there were +1.5 persons to a room; neglect was characterized by high agency

notification of child abuse; a high rate of tuberculosis and infectious disease occurred; high delinquency rates, attempted suicides, and illegitimate births occurred; there was a high birth rate with most of the population under 21; and where housing was subsidized by the government. (6 refs.) - M. Flessinger.

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2500 ROGOW, SALLY. The non-verbal blind child. *New Outlook for the Blind*, 63 (1):1-7, 1969.

A case study of the effects of deprivation on the emotional behavior and language development of a 9-year-old blind girl is presented. Within her family home, she was unable to develop adequate social maturity or expressive language because of the overprotective and defeatist attitude of her mother. However, in the warm, accepting environment of a foster home, she was able to make progress in both areas. Underestimating the potential of a blind child can be a self-fulfilling prophecy. (6 refs.) - J. M. Gardner.

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2501 GARDINER, PETER, MacKEITH, RONALD, & SMITH, VERNON, eds. *Aspects of Developmental & Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 128 p. \$6.00.

At a symposium on pediatric ophthalmology, solutions were discussed for problems which affect children with defective hearing and vision, those associated with special schools, handicapped children, education, and vision testing. Parents of children who fail visual screening tests should contact the hospital eye service, supplementary ophthalmic services, or private doctors. Ophthalmologists should visit special schools in order to see children in their daily routine before the eye examination is given. Specialized provisions should be made for children with defective hearing, sight, and intelligence, because these children present extreme educational and social problems. Small educational units for the handicapped should provide trained teachers, assessment clinics, medical help, and aid to parents. (181 refs.)
V. G. Votano.

CONTENTS: Development of Visual Perception (Vernon); Neurophysiology and Assessment of

Visual Function in Children (Ffoos); The Eye and Vision in the Newborn Infant (MacKeith); Eye Movements and Perceptual Development (Abercrombie); Psychological Aspects of Visual Perception (Tyson); Refractive Errors in Relation to Physical Characteristics and Systemic Disorders (Gardiner); The incidence of Squint in Systemic Disease (Nutt); Vision Screening Procedures for Very Young or Handicapped Children (Sheridan); Electro-oculographic Studies in Cerebral Palsied and in Normal Children (Jones, Dayton, Limpaecher, Murphy, & Hirsch); The "At Risk" Concept with Reference to Visual Disorders (Gardiner); The Diagnosis and Management of Squint (Stanworth); A Screening Service for the Early Detection of Visual Handicap (Smith); Some Developmental Eye Disorders Present at Birth and in the Neonatal Period (Wybar & Harcourt); Children of Reduced Birth Weight (Douglas); Congenital and Early Infantile Cataract (Abrams); Metabolic Disorders Affecting Vision (Gordon); The Eye in Cystine Storage Disease with a Note on the Use of the Slit Lamp (Douglas); The Ophthalmological Examination of the Handicapped Child (Smith); Medical Aspects of the Education of Visually Handicapped Children (Fine); The Conduct of the Interview with the Handicapped Child and his Family (Wigglesworth); Psychiatric Implications of Severe Visual Defect for the Child and for the Parents (Williams); The Education for the Visually Handicapped Child with Additional Disabilities (Longmore); The Education of Children Who are Blind (Brown); Closing Discussion (Gilkes).

2502 GARDINER, PETER. The "at risk" concept with reference to visual disorders In: Gardiner, Peter, MacKeith, Ronald, & Smith, Vernon, eds. *Aspects of Developmental & Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p. 59-61.

Children who are "at risk" of having visual disorders include those born after maternal rubella, those with cyanotic heart disease (75% of these children are "at risk"), those with brain damage (60%), and mongoloid children (80%). Preschoolers and boarding school children should also be considered "at risk." A register for these children should be developed and should include children with familial histories of lazy or crossed eyes. Most eye disorders are discovered only after development has been retarded. An "at risk" register could also function as an observational register with third year ophthalmological examinations for selected children. Mass screening in schools is also recommended. (5 refs.) - V. G. Votano.

2503 FINE, SHIRLEY R. Medical aspects of the education of visually handicapped children. In: Gardiner, Peter, MacKeith, Ronald, & Smith, Vernon, eds. *Aspects of Developmental & Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p. 103-105.

In England and Wales, 20% of the blind children and 13% of the partially sighted have low intelligence and may experience epilepsy (8% of blind and 5% of partially

sighted), maladjustment (9% and 7%, respectively), hearing loss (9% and 4%, respectively), and physical disability (20% and 15%). Of 817 blind children, 134 used visual aids, while of 1,374 partially sighted children, 476 did not use any visual aid. In this group of 476 partially sighted, vision disorders included 116 lens disorders, and 25 children with myopia. Children who did wear eyeglasses were seldom supplied with a spare pair. Additions to general care should include physiotherapy, dental care, and case conferences. (No refs.) - V. G. Votano.

MEDICAL ASPECTS--CONVULSIVE DISORDERS

2504 SPILKER, BERT, & YEAGER, CHARLES L. Ten year EEG studies in epileptic children (two cases). *Diseases of the Nervous System*, 30(3):194, 1969.

EEG recordings from 2 epileptic children (one diagnosed as MR, the other as having a chronic brain syndrome of unknown etiology) showed that, over a 10-year period, the abnormalities in tracings correlated with changes in emotional states. The MR S had a medical regimen which varied from no medication to 27 different combinations of anti-convulsant drugs. Although improvement in the EEG was noted when a new drug was initiated, the records would gradually worsen. At the height of the S's EEG abnormality, tantrums, overactivity, and destructiveness were observed. A total of 154 EEGs were made from the age of 5 to the age of 15 years. In the patient with chronic brain syndrome, a series of 153 EEGs were made from age 3 to age 13 years. His behavior often correlated with his EEG record--a spiking focus in the left temporal region when he was overactive and difficult to manage and a generalized paroxysmal activity when he was quiet and amiable. Since temporary pattern improvement following medication changes occurred in this S also, a relation of EEG change to drug intake rather than to drug specificity is suggested. (No refs.) - A. Huffer.

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San Francisco, California 94122

2505 HALTIA, MATTI, KRISTENSSON, KRISTER, & *SOURANDER, PATRICK. Neuropathological studies in three Scandinavian cases of progressive myoclonus epilepsy. *Acta Neurologica Scandinavica*, 45(1):63-77, 1969.

Neuropathological studies on 3 cases of progressive myoclonus epilepsy (PME) showed a severe loss of Purkinje cells, neuronal loss, degeneration in the medial part of the thalamus, and severe degeneration of the peripheral nervous system. Numerous amyloid bodies were found, particularly in the spinal cord. No inclusions of Lafora bodies in nerve cells were found; however, the 3 cases examined could not be differentiated clinically from PME with Lafora body inclusions. (38 refs.)
L. S. Ho.

*Neuropatologiska Laboratoriet
Patologiska Institutionen I
Sahlgrenska sjukhuset
Goteborg SV, Sweden

2506 GREEN, JOSEPH B. Photosensitive epilepsy: The electroretinogram and visually evoked response. *Archives of Neurology*, 20(2):191-198, 1969.

Sixteen patients with previously demonstrated photosensitive epilepsy were investigated with visually evoked response (VER) electroretinogram (ERG) and conventional EEG in an

attempt to define central and/or peripheral mechanisms in the convulsive response. In 13 patients, the VER was characterized by a large positive response. This tended to be localized over the occipital region, decreased with monocular occlusion, subject to rebound when the eye patch was removed, and augmented by closing the eyes. It was, therefore, related to the phenomenon of vision. In 10 cases, fast flicker-induced EEG seizure activity was associated with an early positive wave in the ERG. It appears that seizure responses in these patients are not mediated by diffuse nonspecific fibers, but rather by specific visual afferents and it is hypothesized that these are failures of inhibitory mechanisms at several levels (retina, lateral geniculate body, cortex), although the site(s) of initial vulnerability have not been established. (14 refs.) - E. L. Rowan.

Department of Neurology
Indiana University Medical Center
Indianapolis, Indiana 46202

2507 FRANTZEN, E., LENNOX-BUCHTHAL, M., & NYGAARD, A. Longitudinal EEG and clinical study of children with febrile convulsions. *Electroencephalography and Clinical Neurophysiology*, 24(3):197-212, 1968.

A total of 218 children whose first convulsion was associated with fever due to extracerebral infection were hospitalized for clinical evaluation and followed with EEGs for up to 5 years. About 1/3 showed "acute" EEG slowing within 6 days after the febrile convulsion. This occurred more frequently in children over 2 years old in whom the convulsion lasted 30 minutes or more, the rectal temperature had been above 39°C, and the illness had lasted for 36 hours or more before the convulsion. Spike and wave abnormalities occurred in only 3 children within 6 hours of convulsing, but were found in 63 within 5 years. Two-thirds of these children had their febrile convulsion after age 2 1/2 years, and those who were younger at the time of initial seizure had a strong family history of convulsions. Twenty-six percent of all children in this series had recurrent febrile convulsions. This was most prominent in the group whose initial seizure had occurred at the age of 13 months or younger. Only 5 children in the entire group developed clinical evidence of "epilepsy." (39 refs.) - E. L. Rowan.

Gentofte Hospital
Copenhagen, Denmark

2508 JENNETT, W. BRYAN. Early traumatic epilepsy: Definition and identity. *Lancet*, 1(7604):1023-1025, 1969.

Three hundred thirty-one patients with non-missile head injuries who had epilepsy in the first 8 weeks after injury were compared with 218 in whom epilepsy was delayed beyond 3 months. Epilepsy began in the first week 29 times as frequently as in any of the subsequent 7 weeks: of patients with epilepsy within 8 weeks of injury, 80.5% had their first fit in the first week. Localized focal motor epilepsy was common in the first week (40%), infrequent in the next 7 weeks (17%), and rare after that (3%). Temporal-lobe epilepsy was never recorded in the first week, but was as common during the next 7 weeks as it was after 3 months (20%). First-week epilepsy recurred or persisted in only 27% of patients, but from the second week onwards, the recurrence-rate was over 70%. A small missile series (73 patients) showed similar trends in regard to all the above factors. Early epilepsy has, therefore, distinctive characteristics which justify separate classification, but the term should be confined to fits in the first week after injury. Although less than 1/3 of patients with early epilepsy have further fits in the future, this risk is significantly greater than for those without early epilepsy, which, therefore, has prognostic significance. (11 refs.) - *Journal summary*.

Institute of Neurological Sciences
Glasgow, Scotland

2509 LIVINGSTON, SAMUEL. When to hospitalize the epileptic child. *Hospital Practice*, 4(3):77-79, 84-86, 1969.

Epilepsy in its multiple forms is best managed conservatively on an ambulatory basis, but there are occasions when hospitalization is necessary for patient safety or control. Prolonged seizure activity is a medical emergency and requires vigorous treatment. Supervised drug administration may be necessary to differentiate patient unreliability from a metabolic abnormality. A change in environment may relieve some seizures associated with emotional conditioning. Extensive workups may be necessary to investigate neurological abnormalities or symptom changes or to support surgical intervention. The initiation of unconventional starting doses of anticonvulsive medication or the institution of a ketogenic diet is best done in the

hospital. Inpatient observation may be necessary in order to differentiate epileptic seizures from hysterical symptoms or convulsions associated with breath-holding spells. (No refs.) - E. L. Rowan.

Johns Hopkins University School of
Medicine
Baltimore, Maryland 21205

2510 McINERNEY, THOMAS K., & *SCHUBERT, WILLIAM K. Prognosis of neonatal seizures. *American Journal of Diseases of Children*, 117 (3):261-264, 1969.

Among 95 infants in whom seizures were noted during the first 30 days of life, 70 were followed-up by review of hospital records or contact with family physician or parents. Forty percent of the group were reported as normal, 20% died, and 34% was abnormal (27% MR). The etiology of the convulsions was most significant in the prognosis. The hypocalcemic group did very well, while those with perinatal complications (birth trauma) did poorly. Patients with congenital anomalies and central nervous system infections had the highest death rate while those with hypoglycemia and seizures of unknown etiology had a mixed prognosis. Although few EEGs were done in this series, they confirmed earlier findings that EEG abnormalities of any kind were associated with poor prognosis. Of some value in counseling parents of infants with seizures, these findings should also challenge the physician to seek to prevent birth trauma and its unfortunate sequelae. (7 refs.) - E. L. Rowan.

*Children's Hospital Research Foundation
Elland Avenue and Bethesda
Cincinnati, Ohio 45229

2511 RODIN, ERNST A. *The Prognosis of Patients with Epilepsy*. Springfield, Illinois, Charles C. Thomas, 1968, 455 p., \$19.50.

Most investigators agree that only 1/3 of patients treated for seizure disorders will have remission of symptoms for at least 2 years. In his initial evaluation of the epileptic patient, the physician would like to predict such a prognosis. Computer analysis of several variables in the life history of epileptics has delineated several significant factors. Long duration of untreated illness, combinations of seizure types (especially psychomotor seizures), and a high frequency

of seizures are highly correlated with a poor prognosis, while early onset of symptoms and the degree of EEG abnormality are of less prognostic value. It is important to recognize patients with "cerebral damage" as this is associated with low intelligence and a poor prognosis. Seizures themselves result in a minimal lowering of tested intelligence which may be dramatic only in individuals with initially high levels of function. It appears extremely important to effect early and complete seizure control, if possible--as this tends to prevent permanent sequelae. In the future, more attention must be paid to constitutional factors and statistically controlled follow-up. Undue optimism is deadly and both practicing physicians and researchers must respect epilepsy and its consequences. (309 refs.) - E. L. Rowan.

CONTENTS: Results of Follow-up Studies and General Prognostic Criteria; Childhood Epilepsy; Febrile Convulsions; Petit Mal; Infantile Spasms--Hypsarrhythmia, Akinetic and Myoclonic Seizures; Posttraumatic Epilepsy; Surgery; Intelligence; Mortality; Introduction; Prognosis for Seizure Control; Prognosis for Behavior; Prognosis for Intellectual Functions; Prognosis for Employment; Institutionalization; Life Expectancy; Summary.

2512 PRYSE-PHILLIPS, WILLIAM. *Epilepsy*. Bristol, England, John Wright & Sons, 1969, 96 p. \$2.06.

Epilepsy in its various forms is reviewed with special emphasis on etiology, clinical features, drug therapy, EEG, and social and psychiatric management. A person with epilepsy is seen as an individual in an unenlightened society where his disability is reinforced. The physician has a duty to be properly informed about epilepsy and not only treat his patient physically, but also aid his integration into society. A practical classification scheme (centrencephalic and focal) is given extensive clinical and physiological documentation. This book presents an excellent clinical review for the physician, and although directed at the British practitioner, it might be adapted easily in other countries. (12-item bibliog.) E. L. Rowan.

CONTENTS: Discussion and History; Neurophysiology; Synthesis: A Classification; Aetiology and Incidence; The Clinical Features of an Attack (I); The Clinical Features of an Attack (II); The E.E.G. in Diagnosis; Drug Therapy; Social Management; Psychiatric Management; Survey.

- 2513 GRASSET, ALBERT. *L'enfant Epileptique (The Epileptic Infant)*. Paris, France, Presses Universitaires de France, 1968, 252 p., \$1.81.

Epilepsy in children is treated as a problem of medicine, psychiatry, and social sciences. The first part of this book describes the symptoms, etiology, diagnosis, and treatment of epilepsy. The second part discusses the relations of an epileptic child with his physician, family, and school, and the prognosis for living a relatively normal life. Physicians, physiologists, psychiatrists, and social scientists should be interested in this book. (140 refs.) - L. S. Ho.

CONTENTS: Symptoms and Phenomena of Epilepsy; Epileptics and Epilepsies: Time and Space Distributions of the Symptoms of Epilepsy; Symptoms to be Observed in Young Epileptics; Positive Diagnosis of Epilepsy; Differential Diagnosis of the Types of Epilepsy; Factors Pre-disposing to Convulsions and Epilepsy; Respective Roles of Heredity Versus Environment in the Etiology of Epilepsy; Treatment of Epilepsy; The Epileptic Child and his Doctor; The Epileptic Child and his Family; The Epileptic Child and his School; Recreation; Professional Work; Driving an Automobile; Marriage; Prospectives for Living a Normal Life.

- 2514 WHISLER, K. E., TEWS, J. K., & STONE, W. E. Cerebral amino acids and lipids in drug-induced status epilepticus. *Journal of Neurochemistry*, 15(2):215-220, 1968.

Cerebral tissue frozen *in situ* was removed from dogs after 30-40 minutes in pentylenetetrazol-induced status epilepticus and analyzed for amino acids and lipids. Levels of alanine, arginine, γ -aminobutyric acid, glycine, histidine, leucine, lysine, phenylalanine, serine, tyrosine, and valine showed significant increases, while those of glutamic and aspartic acids were decreased. The ganglioside fraction and a fraction containing the lecithins and sphingomyelins were decreased, while the cephalin, cerebroside-sulphatide, and cholesterol fractions were not changed. Shorter seizures induced with bemegride brought about less extensive changes with increases in glutamic acid, alanine, γ -aminobutyric acid, lysine, and ammonia and decreases in the ganglioside and lecithin-sphingomyelin fractions. Some of the changes noted may be related to decreased cerebral oxygen tension which is known to decrease during convulsive activity. It is

also possible that the seizure alters the rates of entry or exit of amino acids. (36 refs.) - M. G. Conant.

Department of Physiology
University of Wisconsin Medical School
Madison, Wisconsin 53706

- 2515 McLARDY, TURNER. Ammonshorn pathology and epileptic dyscontrol. *Nature*, 221 (5183):877-878, 1969. (Letter)

Ammonshorn sclerosis as classically found in the brains of epileptics has been reproduced experimentally in guinea pigs with repetitive seizures in a hypoxic, hyperthermic state. This may be analogous to anoxia during febrile convulsions in childhood. Subsequent epileptic dyscontrol may be the result of abnormal activation of the amygdaloid by the sclerosed ammonshorn, and leukotomy through these connections may cure the seizures and dyscontrolled behavior. (6 refs.) - E. L. Rowan.

Myerson Research Laboratory
Boston State Hospital
Boston, Massachusetts 02124

- 2516 WECKMAN, NILS, & LEHTOVAARA, RAIMO. Folic acid and anticonvulsants. *Lancet*, 1(7587):207-208, 1969. (Letter)

The concentration of folate in cerebrospinal fluid was found to be unrelated to epilepsy, anticonvulsant medication, or coincident neuropsychiatric abnormalities. In addition, there were no consistently low serum folate values in epileptics with or without anticonvulsants. A causal relationship between long-term medication and disturbance of folate metabolism has not been demonstrated. (15 refs.) - E. L. Rowan.

Department of Neurology
University of Helsinki
Helsinki, Finland

- 2517 GORDON, NEIL. Use of diphenylhydantoin in epilepsy treatment. *Developmental Medicine and Child Neurology*, 11(1):111-112, 1969.

Drug treatment for epilepsy with diphenylhydantoin is complicated by the problem of toxic side effects. Even with average doses of 300 mg/day (or 4 mg/kg body weight), neurological complications can occur. Early

signs include nystagmus, blurred vision, diplopia, and dizziness. Severe reactions are rare, although certain forms of epilepsy (particularly petit mal and myoclonic seizures) may become worse. The greater use of laboratories to establish therapeutic serum concentrations would avoid the toxic effects. (11 refs.) - J. M. Gardner.

Booth Hall Children's Hospital
Charlestown Road
Blackley, Manchester 9, England

2518 VIUKARI, N. M. A. Diphenylhydantoin as an anticonvulsant. Evaluation of treatment in forty mentally subnormal epileptics. *Journal of Mental Deficiency Research*, 13(3):212-218, 1969.

Anticonvulsant treatment of SMR epileptic Ss with diphenylhydantoin sodium (Dilantin) with

or without tranquilizers is very effective. The ability of Dilantin to suppress seizures in MR epileptics in whom other sedatives might have caused psychomotor dysfunction was studied. Forty grand mal, focal, and psychomotor epileptic Ss were withdrawn from other drugs and given Dilantin over a 1- to 2-month period. Seizure frequency was checked 6 months prior to and after the changeover. The Ss were maintained on strict diets 2 weeks before and during the trial. Dilantin treatment decreased seizure frequency in 20 cases, increased it in 4, and had no effect in 16. Those Ss changing from sedatives to Dilantin became more mentally alert and became better at self-care. Intensive and controlled treatment with one anticonvulsant is more effective than random use of different compounds. (19 refs.) - R. K. Butler.

Research Department
Rinneke Institute for The Mentally Retarded
Majalampi, Finland

MEDICAL ASPECTS--CHROMOSOMAL

2519 MOTULSKY, ARNO G. Population genetics of mental retardation. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 3, p. 13-21.

The development of the 1/2 to 3/4 of man's intelligence which appears to be determined by genetic factors seems to be related to the undefined physiologic action of multiple interacting polygenes. Mild MR can be caused by normal quantitative variation in these polygenes, sex chromosome anomalies, a sub-cultural environment, or a cerebral disease in childhood. About 10% of SMR, exclusive of trisomy and identifiable exogenous causes, is caused by autosomal recessive genes, and the etiology of the other 90% is unknown. Mutation pressure may be responsible for the retention of recessive genes for MR in the population. Predisposition to chromosomal anomalies may be related to autoimmunity and structural chromosome rearrangement. Mild MR is prevalent among the lower social classes, while SMR is found in all social classes. Although individuals with mild MR have a higher rate of reproduction than normals, the intelligence of the population does not decrease because the high fertility rate of married MRs is counterbalanced by a high proportion of childless siblings. (28 refs.) J. K. Wyatt.

2520 BOOK, J. A. Geographical isolates in the genetics of mental retardation. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 1, p. 5-8.

Genetic factors which are important causes of MR are: the minus variants of a normal distribution resulting from the interaction of polygenes and the environment; specific major gene mutations; and chromosome mutations. Most knowledge of rare recessive disorders has come from studies of geographically isolated populations. These have generally shown that small numbers of ancestral pairs from 5 to 7 generations earlier were responsible for most of the present population. In an investigation of low-grade MR in 3 parishes in Sweden which covered the period from 1900 to 1930, 180 patients belonging to 134 families were discovered. Of these, 141 (91 males, 50 females) belonged to 102 families, all of whom had descended from a few ancestral pairs in the middle of the seventeenth century. Their MR was congenital and stationary and appeared to be due to a simple recessive type of inheritance. Although speech was dysarthric and many Ss had pithicoid posture, physical development was remarkably normal, skulls were well-shaped, none had malformations, and neurological examinations were normal. In a follow-up

study of 2 parishes in 1960, 318 propositi and secondary cases of MR were identified. Approximately 202 of these were etiogenetically identical, and their MR and other signs and symptoms were mainly due to homozygosity for a major recessive mutation. These findings indicate that there are major genic and polygenic causes in the region of overlapping IQs for those MRs who remain after MRs with recognizable specific diagnoses have been excluded. (6 refs.) - J. K. Wyatt.

- 2521 LENZ, WIDUKIND. Prospects in prevention of trisomic conditions based on maternal age. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 2, p. 9-12.

The basis of trisomy is nondisjunction, its etiology is unknown, and most cases occur as unpredictable, sporadic events. Maternal age is the only factor which has a straightforward relation to Down's syndrome. The incidence of this syndrome increases slightly until a maternal age of 30 years and more than doubles every 5 years after age 30. The relative increase in risk for trisomies 21, D, E, and X and for XXY-Klinefelter's syndrome is similar to that for Down's syndrome. If the reproductive age in women were lowered, the incidence of mongolism and of other trisomies associated with MR and malformations would be appreciably reduced. Evidence that dominant gene mutations such as achondroplasia and actrocephalosyndactyly demonstrate increased frequency with increased paternal age suggests that a reduction in parental reproductive age would be a successful preventive measure. (No refs.) - J. K. Wyatt.

- 2522 ARAKAKI, DAVID T., WAXMAN, SORRELL H., NONOMURA, TULINE M. Anomalies of chromosome No. 3 in abortions. *Journal of Medical Genetics*, 6(4):399-400, 1969.

A ring chromosome number 3 was found in an early spontaneous abortion. The clinical data of 2 additional trisomy-3 and a monosomy-3 were reviewed. Abnormalities of chromosome number 3 are apparently lethal in very early embryonic development. (6 refs.) *Journal summary*.

Department of Genetics
School of Medicine
University of Hawaii

- 2523 BREG, W. ROY. Cri du chat syndrome. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 9, p. 632-638.

The clinical features of *cri du chat* syndrome include a peculiar cry in infancy, growth failure, craniofacial abnormalities, microencephaly, simian creases, and psychomotor retardation and MR in patients old enough to be tested. The deletion of a B-group chromosome, usually number 5, is presumably the cause of the phenotypic abnormalities. The etiology of deletion is unknown. In a few cases the deletion is inherited from phenotypically normal balanced translocation carriers. Since the patients are often severely MR (IQ around 20), their management is rather difficult. (20 refs.) - L. S. Ho.

- 2524 GELLIS, SYDNEY S., & FEINGOLD, MURRAY. Cri du chat syndrome (Cry of the cat syndrome). *American Journal of Diseases of Children*, 117(6):699-700, 1969.

Five pictures of children with the *cri-du-chat* syndrome are presented. Manifestations of this syndrome include SMR, microcephaly, rounded facies, ocular hypertelorism and a cat-like cry. There is a deletion of chromosomal material from the number 5 chromosome. Institutional care is usually required. (4 refs.) - F. J. McNulty.

Boston Floating Hospital
20 Ash Street
Boston, Massachusetts, 02111

- 2525 NEUMAN, ITTAI, LEVIN, STANLEY, & STRICHOVSKY, PAUL. "Cri du chat" syndrome. *Harefuah*, 74(4):1-3, 1968.

A 2-year-old female exhibited the criteria of the *cri-du-chat* syndrome including: a peculiar cry, MR, physical retardation, microcephaly, hypertelorism, antimongoloid eye slant, epicanthal folds, low-set ears, hypognathia, and generalized hypotonia. Chromosomal analysis revealed that the fifth chromosome had a partial deletion on the short arm. Abnormal dermatoglyphic features included a triradius and loop (no whorl) on each finger and a 57° *abd* angle on the left hand. The child had normal reflexes. (6 refs.) - A. Huffer.

Pediatrics Department
Kaplan Hospital
Rehovot, Western Galilee

2526 HIGURASHI, MAKOTO, NAGANUMA, MASUYOSHI, MATSUI, ICHIRO, & KAMOSHITA, SHIGEHICO. Two cases of trisomy C6-12 mosaicism with multiple congenital malformations. *Journal of Medical Genetics*, 6(4):429-434, 1969.

Two infants with multiple congenital malformations, including abnormally-shaped skull, a peculiar appearance, short neck, long and slender trunk, abnormal skin, and SMR, are reported. Agenesis of the corpus callosum was noted in one case by pneumoencephalogram. Chromosomal analysis in these 2 cases showed 46,XX/47,XXC+ and 46,XY/47,XYC+. The extra chromosome in 47 chromosome cells was shown to be a C group autosome and not an X chromosome by means of autoradiography. (12 refs.) *Journal summary*.

Department of Pediatrics
University of Tokyo
Tokyo, Japan

2527 KELLY, SALLY, ALMY, RYDIA, & DAGLE, ALICE. Chromosome 15 abnormality in a mentally retarded adult. *Journal of Medical Genetics*, 6(4):438-441, 1969.

A small, submetacentric, forty-seventh chromosome was found in the karyotype of an MR woman with skeletal abnormalities. A D₁₅ chromosome was missing from the karyotype of her normal brother and was apparently replaced by a similar, small, unmatched chromosome. The pedigree included a retarded nephew and evidence of transmission of a balanced translocation through the maternal line. The clinical picture, dermatoglyphs, and indices of hematological, immunological, and biochemical maturity differed from those of the D₁ (13) trisomy. (20 refs.) - *Journal Summary*.

Birth Defects Institute
New York State Department of Health
Albany, New York

2528 GREEN, J. R., JR., KROVETZ, L. J., & TAYLOR, W. J. Two generations of 13-15 chromosomal mosaicism: possible evidence for a genetic defect in the control of chromosomal replication. *Cytogenetics*, 7(4):286-297, 1968.

An identical nontranslocation autosomal mosaicism was noted in a phenotypically normal mother and 2 phenotypically abnormal daughters. One daughter has a dull personality, minimal facial and skeletal abnormalities, and mild congenital heart disease

while the other has severe MR, impaired physical growth, moderately severe congenital heart disease, and many skeletal abnormalities. Two other siblings are normal. All 3 affected persons were demonstrated to be 46/47 mosaics with the extra autosome in the 13-35, or D, group. A dominant mutation may have resulted in defective control of the mechanisms which regulate DNA replication of a specific 13-15 group chromosome, and the mutant gene was probably passed by the mother to 2 of her 4 children, thus explaining the inheritance of somatic mosaicism in this family. (45 refs.) - M. G. Conant.

Department of Medicine
University of Florida
Gainesville, Florida 32603

2529 JUBERG, RICHARD C., ADAMS, MORTON S., VENEMA, WILLIAM J., & HART, MERRILY G. Multiple congenital anomalies associated with a ring-D chromosome. *Journal of Medical Genetics*, 6(3):314-321, 1969.

A female infant with microcephalus, hypertelorism, eye and ear anomalies, epicanthi, micrognathia, short neck, hypoplastic nipples, congenital heart disease and hip dysplasia, absent thumbs, simian creases, feet anomalies, and mental and physical retardation was determined to have a ring-D karyotype. A similar constellation of symptoms had been noted in a stillborn infant and a 5-year-old child. Since a ring chromosome is formed by the loss of chromatin material at both ends, the deletion may explain the defects found in these 3 cases. Although other cases of D-ring chromosome anomalies are dissimilar to these 3 cases, it is possible that the deleted material was translocated partially to other chromosomes, thus changing the phenotype. It is considered unlikely that this rare chromosomal aberration and the constant phenotypes in the 3 cases noted are accidental. (40 refs.) - E. Hays.

University of Michigan Medical School
Ann Arbor, Michigan 48104

2530 VARELA, MARIA A., & STERNBERG, WILLIAM H. Ring chromosomes in two infants with congenital malformations. *Journal of Medical Genetics*, 6(3):334-341, 1969.

Two cases of ring chromosomes are reported; in one child the ring replaced a chromosome of the D group (13-15), and in the other S, a ring chromosome and a mosaic trisomy state were found. One infant had low-set protruding ears, "cafe au lait" spots, episodes of

respiratory infections, abnormal dermatoglyphics, interventricular and interatrial defects, and hypertrophy of the adrenal medulla. She died at 15 months of age. The mother had no history of consanguinity nor irradiation. The second child was a mosaic with one line consisting of normal 46,XY plus a ring chromosome of unknown origin. This infant had abnormal ear lobes, abnormal dermatoglyphics, polythelia, unilateral cryptorchidism, micrognathia, hypertelorism, and congenital heart disease. This child's mother had no history of irradiation nor consanguinity. The identity of the ring could not be established from the data; however, the congenital malformations suggest a probable derivation from an autosome. Recent chromosome studies of peripheral blood leukocytes of this patient at 18 months showed no difference in the percentage of cells which contained the ring, although in a similar case, instability and eventual loss of the ring occurred over a period of 13 months. (24 refs.) - L. E. Bayliss.

Tulane University School of Medicine
New Orleans, Louisiana 70112

2531 DE CAPOA, A., BREG, W. R., KUSHNICK, T. & MILLER, O. J. Radioautographic identification of the D chromosomes involved in the centric fusion type of D/G translocation, t(DqGq). *Annals of Human Genetics*, 32(2): 191-193, 1968.

Radioautographic studies of 5 patients with a D/G translocation determined that the D group chromosome number 14 was involved in 3 cases and D group chromosome number 15 in the remaining 2. Four unrelated D/G translocation mongoloids, in an institution for the MR, and one unrelated normal carrier were studied. Family history for mongolism was negative in 2 or more generations for the 4 mongoloid patients, but the normal carrier had a mongoloid brother. Results of this study were compared with previous data indicating that in a series of D/G translocations, the D-group chromosome involved is either number 14 or 15, with number 14 most frequently involved (8 refs.) - B. Bradley.

Columbia University
New York, New York 10032

2532 SMITH, DAVID W. The 18-trisomy and D₁-trisomy syndromes. In: Gardner, Lytt, I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 9, p. 638-652.

MR is the most prominent feature in trisomy 18, and D₁. Both chromosomal abnormalities are relatively rare in newborns and the majority of these infants die before the third month of life. Chromosome studies should be done on the baby and also on the parents if the baby is a translocation partial trisomy case. If either one of the parents is a carrier, the risk of having another affected child would be significant. The most common clinical features of trisomy-18 are altered skeletal proportions, hypotonia, unusual positioning of the clenched fingers, micrognathia, MR, and growth retardation. Some of the features also occur in E-trisomy, D₁-trisomy, and the Smith-Lemli-Opitz syndrome. The common characteristics of D₁-trisomy are cleft lip and cleft palate plus microphthalmia with colobomata and forebrain hypoplasia. The diagnosis of D₁- or 18-trisomy should be confirmed by cytogenetic studies. (44 refs.) L. S. Ho.

2533 MIKELSAAR, A. -V. N., & TALVIK, T. A. Partial deletion of the long arm of chromosome 18. *Humangenetik*, 7(4):359-360, 1969.

A female infant who had a partial deletion of the long arm of chromosome 18 exhibited SMR and multiple physical anomalies. The proband was the product of the third pregnancy--the first ended in a spontaneous abortion, the second terminated at the seventh month with a stillborn male infant. Somatic anomalies included plagiocephaly, visible forehead veins, short neck, low-set and abnormal ears, asymmetrical face, a left eye with coloboma and chorioidea, unequal eye size, a flat nasal bridge, small nose, "carp" mouth, genu valgum, flat feet, abnormally located toes, 4 whorls on the fingers, underdeveloped labia minores, muscle hypotonia, and a peculiar cry. The father had a normal chromosome complement; however, the mother, who also had chronic blepharitis, exhibited chromosomal aberrations (markers, dicentrics, rings, and fragments) in 25 to 70% of the cells. It may be that the proband's chromosomal deletion has a viral etiology. (11 refs.) - A. Huffer.

Karyological Laboratory
Vavilov str. 32
Moscow B-312, Union of Soviet Socialist Republic

- 2534 HECHT, FREDERICK. IgA and partial deletions of chromosome 18. *Lancet*, 1(7585):100-101, 1969. (Letter)

A 5-year-old SMR, female dwarf with the karyotype 47, 18+ has immunoglobulin (Ig) deficiencies (IgA-severe, IgM-moderate, and IgG-mild) and κ and λ light-chains deficiencies. In the light of these findings, the mapping of the locus for IgA on 18q is challenged. (8 refs.) - A. Huffer.

Division of Medical Genetics
University of Oregon Medical School
Portland, Oregon 97201

- 2535 PENROSE, L. S. Dermatoglyphics in trisomy 17 or 18. *Journal of Mental Deficiency Research*, 13(1):44-59, 1969.

Analyses of the dermatoglyphics of 22 cases of trisomy 17 or 18 found, in addition to a marked frequency of arch formations, a tendency for displacement distally of the tri-radius t on the palms. The soles of the feet were characterized by presence of e or p tri-radii with absence of f associated with one or more distal loops on areas II, III, and IV. Studies were made on 6 males and 16 females with trisomy 17 or 18, on 2 incomplete cases, and on 4 cases with deletion of chromosome 17 or 18. Among the Ss with trisomy 17 or 18 (a total of 160 fingers), there were 86.9% with arch formation, 10% with annular loop, 1.9% with a whorl, and 1.2% with a radial loop. In cases of deletions of 17 or 18, whorls predominated, and arches were absent. In these cases, the frequency of whorls exceeded that in normal controls by 18.9%. The examination of palms gave somewhat equivocal findings in that 9 cases could be considered normal, 4 had very unusual patterns with distally placed axial tri-radii t , and on 10 of 34 palms, the A-line exited at the thenar border while normal expectancy would be 4 of 34. Toe patterns were similar to fingers. Soles were variable with the most notable feature being reduction of total number of loops. Although the trisomy was not further distinguished than 17 or 18, it was believed that 18 was the usual chromosome involved. A trend was noted among the parents to a higher mean age for both father and mother. (11 refs.) - C. A. Rizvi.

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Harperbury Hospital
St. Albans, Hertfordshire, England

- 2536 JENSEN, KNUD, CHRISTENSEN, KIRSTEN, RIBER, JACOBSEN, PETREA, NIELSEN, JOHANNES, FRIEDRICH, URSULA, & TSUBOI, TAKAYUKI. Ring chromosome 18 and gamma-M-globulin abnormality. *Lancet*, 2(7618):497-498, 1969.

Serum immunoglobulin studies performed on an MR mother and daughter, both with ring chromosome 18, revealed that the mother had a serum γ -M-paraprotein and increased total γ -M-globulins. Protein studies in the daughter were normal. (10 refs.) - E. L. Rowan.

Arhus State Hospital
Risskov, Denmark

- 2537 HADDAD, ZACK H., ALLEN, RAY F., TOWNER, JOSEPH W., & WILSON, MIRIAM G. IgA, IgM, and partial deletion of chromosome 18. *Lancet*, 1(7596):678, 1969. (Letter)

Of 4 patients with chromosomal abnormalities (2 with 18q-; 1 with 4p-; 1 with a ring D), one patient with deletion of the long arm of chromosome 18 demonstrated a persistently low level of immunoglobulin (Ig) M for his age (7 mg/100 ml at 2 yrs 9 mos; 17 mg/100 ml at 3 yrs 3 mos) but a normal level of IgA and IgG. The other 2 Ss had normal IgA, IgG, and IgM values. In reporting Ig levels, the values should be interpreted in terms of mean values for age \pm 2 standard deviations. (10 refs.) - A. Huffer.

Department of Pediatrics
Southern California Medical Center
Los Angeles, California 90033

- 2538 SINHA, ANIL K. Human ring chromosome syndromes: An "E" ring associated with an abnormal phenotype. *Acta Geneticae Medicae et Gemellologiae*, 27(3):487-493, 1968.

Clinical examination of a 16-month-old female revealed apathy, slow response to stimuli, retarded verbal and psychomotor development, epicanthal folds, bilateral dual palmar creases, and extremely short fifth fingers. The cranial nerves were intact, reflexes were normal, laboratory data was within normal limits, and there was no evidence of congenital glaucoma. In peripheral blood cultures, a ring-shaped chromosome was observed in place of an E-group member in 100% of 25 cells. The other chromosomes did not show any obvious structural anomalies, and the ring chromosome maintained a fairly constant size in individual metaphases. Although the parents and male sibling of the patient were unavailable for chromosomal examination, it

was suggested that this ring chromosome has become a permanent part of the patient's chromosome complement. (11 refs.)

M. G. Conant.

Baylor University College of Medicine
Texas Medical Center
Houston, Texas 77025

2539 HELLER, MAX. Die Mongoloidie in der Geschichte und als heilpädagogische Aufgabe (Mongolism in history and as a problem of medical pedagogy). *Schweizer Erziehung-Rundschau*, 41(9):245-248, 1968.

In examining the knowledge of mongolism from an historical perspective, a pre-scientific era and a scientific era are seen: the former encompassing the time before 1866 when Down published his paper, and the latter falling into 2 phases characterized by clinical observations of phenomena until 1959, and etiological studies since the discovery of the chromosomal abnormality underlying the syndrome. In the prescientific era, no written work is noted before 1800; however, in paintings of the fifteenth and seventeenth centuries, at least 3 known examples of mongoloid types exist: 2 Madonnas with mongoloid Christ-child by Mantegna (1431-1506) and a work by Jordaens (1593-1678). Before 1800, mental illness, then feeble-mindedness and around 1840, cretinism were incriminated, but in 1866 investigations of mongolism with exact description of internal and external characteristics began. Down's article led the way and, until 1900, most work was done in England. German research was delayed because the syndrome had been considered a form of cretinism or congenital rickets. From 1902 to 1959, new observations and theories including chromosomal abnormalities emerged throughout the world. When the normal count was proven to be 46 instead of 48, the way was opened for the work which proved trisomy 21 to underlie the clinical syndrome. Four forms are now recognized: true trisomy 21; translocations; mosaicism; and others with chromosome counts ranging from 47-50. If the future can be projected in terms of the past, it is estimated that by 2040 prophylaxis will be effectively preventing mongolism. Meanwhile, the problems of health and life-span in mongoloid populations must be dealt with. (7 refs.) - C. A. Rizvi.

No address

2540 BREG, W. ROY. Mongolism (Down's syndrome). In: Gardner, Lytt, I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 9, p. 608-631.

Mongoloid facies and MR are the 2 constant features of Down's syndrome, although MR is not always apparent in infancy. No specific preventive or corrective therapies for the general physical or mental abnormalities of Down's syndrome are available. Medical care for these patients is essentially the same as for normal Ss. The overall incidence of Down's syndrome is 1/600 births and increases with maternal age from 0.6/1,000 at maternal ages of less than 30 years to about 20/1,000 for mothers 45-years-old or over. The recurrent risk rate for giving birth to a mongoloid is appreciably increased for mothers under 30 years, but is almost unaffected for mothers over 30 years. The clinical and radiologic manifestations, diagnosis, associated complications, and cytogenetics of Down's syndrome are reviewed. (110 refs.)

L. S. Ho.

2541 POLANI, P. E. Cytogenetics of Down's syndrome. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 4, p. 22-31.

The major cytological variants of Down's syndrome in males are primary trisomy 21, double primary trisomies (21 and XXY), interchange trisomies (D/21 and G/21), and mixoploidy. There is a high incidence of double trisomy at birth (1/16 in newborn males with Down's syndrome). Studies of 20 families in which 2 different aneuploid conditions affected different siblings indicate that the distribution of chromosomal aneuploidies may not be random in some families. The frequency of the interchanges in Down's syndrome vary with maternal age. About 10% of patients with mothers under 30 and approximately 1.5% of patients with mothers over 30 have interchange. The overall proportion of mixoploidy in cases of Down's syndrome is 2.4%. In mixoploidy and incomplete trisomy, cellular events and cell-selection pressure may gradually change the chromosome composition of tissues sample for chromosome studies. (62 refs.) - J. K. Wyatt.

- 2542 KASHGARIAN, MARK, & RENDTORFF, ROBERT C. Incidence of Down's syndrome in American Negroes. *Journal of Pediatrics*, 74(3):468-472, 1969.

Data relating to the incidence of children with Down's syndrome in Memphis-Shelby County, Tennessee for an 11-year period indicate no difference between rates of this syndrome for Caucasian and Negro populations. Birth records from 5 major hospitals in the locality were surveyed as well as data from pediatricians, nurses, and other agencies. Rates for Negro populations were 1.02/1,000 live-births with 92 infants identified as having Down's syndrome. There were 100 infants with Down's syndrome for the Caucasian population and a rate of 0.95/1,000 live-births. This syndrome occurred for both groups in "time clusters" which could be an indication of an epidemic environmental factor in etiology of chromosomal anomalies. (12 refs.) - B. Bradley.

University of Tennessee
Knoxville, Tennessee 37916

- 2543 HIGURASHI, MAKOTO, MATSUI, ICHIRO, NAKAGOME, YASUO, & NAGANUMA, MASUYOSHI. Down's syndrome: Chromosome analysis in 321 cases in Japan. *Journal of Medical Genetics*, 6(4):401-404, 1969.

The frequency of G21-trisomy and of translocation in Down's syndrome in Japan was evaluated. The frequency of translocation in affected children born to mothers younger than 30 years at parturition was about 7.1%, and the frequency of inherited translocation was at least 2.9% in that group. The frequency and proportions of translocated chromosome were evaluated by means of autoradiography. (8 refs.) - *Journal summary*.

Department of Pediatrics
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Tokyo, Japan

- 2544 MATSUNAGA, E., & MARUYAMA, T. Human sexual behaviour, delayed fertilization and Down's syndrome. *Nature*, 221(5181):642-644, 1969.

An investigation of the hypothesis that the correlation between advancing maternal age

and incidence of Down's syndrome is a function of delayed fertilization due to a decreasing frequency of marital coitus with age revealed that individual variations in frequency of marital coitus for the age groups 21-25, 31-35, 41-45, and 51-55 follows a Poisson distribution. Statistical analysis of sexual behavior for ages 16-60, based on Kinsey's data, revealed that the increase in the expected frequency of nondisjunction with advancing age is slow and that frequency in the oldest group is less than twice that in the youngest group. This analysis was based on assumptions that the probability that the gametes remain fertilizable is a function of time for both the sperm and the oocytes, and that oocytes predisposed to chromosome nondisjunction deteriorate as a function of time. Although the relation between the incidence of Down's syndrome and advancing maternal age has been established, German's delayed fertilization hypothesis does not explain the empirical data unless age-related factors other than prolongation of interval between coitions can be identified. (7 refs.)

J. K. Wyatt.

National Institute of Genetics
Mishima, Japan

- 2545 MARMOL, JOSE G., SCRIGGINS, ALAN L., & VOLLMAN, RUDOLF F. Mothers of mongoloid infants in the collaborative project. *American Journal of Obstetrics and Gynecology*, 104(4):533-543, 1969.

The obstetrical and clinical histories of mothers of 61 mongoloid infants were compared with those of control mothers matched for age, race, hospital, and date of last menstrual period. The incidence of mongolism in Negroes and whites was nearly the same. There was a bimodal distribution of maternal age with one peak at age group 20-24 (largely white mothers) and a second peak at age group 40-44 (largely by Negro mothers). There were no differences in age at menarche, parity, prior abortions, birth order, or pregnancy-free interval between mothers of mongoloids and controls. Both groups reported the same low incidence of organic disease, drug use, and X-ray procedures. Mongoloid infants had a one-week shorter period of gestation and were an average of 200 gm lighter than control infants. Fifty-seven percent of the mongoloid infants were males. (39 refs.)

E. L. Rowan.

No address

2546 CASTALDO, VINCENZO. Down's syndrome: A study of sleep patterns related to level of mental retardation. *American Journal of Mental Deficiency*, 74(2):187-190, 1970.

EEG, EMG, and EOG were continuously recorded in 10 adolescent mongoloid male Ss who slept 4 consecutive nights in the laboratory. Ss were divided into 2 groups matched for age: moderate and severe MR. The severe group had less rapid eye movement (REM) sleep time and greater REM-latency than the moderate group. The discussion of these results referred to the possible relationship between REM-sleep and intellectual functioning, to the psychoanalytic theory of dreams, and to recent biochemical discoveries. (17 refs.) - *Journal abstract*.

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Albany, New York 12208

2547 KEELE, DOMAN K., RICHARDS, CONSTANCE, BROWN, JAMES, & MARSHALL, JANE. Catecholamine metabolism in Down's syndrome. *American Journal of Mental Deficiency*, 74(1):125-129, 1969.

Ss with Down's syndrome (DS) have been demonstrated to have abnormal serotonin metabolism. The purpose of this investigation was to study the metabolism of another group of monoamines, the catecholamines, in this condition. The Ss were paired with MR, non-DS patients as control Ss, and catecholamine and catecholamine metabolite determinations were done on urine; catecholamine determinations of the plasma were also made. Urinary dopamine, 3 methoxy--4-hydroxymandelic acid, 3,4-dihydroxymandelic acid, metanephrine, normetanephrine, and norepinephrine levels were not statistically different from controls. Urinary epinephrine levels were statistically lower than controls. Plasma epinephrine and norepinephrine levels were the same as controls. (19 refs.) - *Journal abstract*.

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2548 APPLETON, M. D., HAAB, W., BURTI, U., & ORSULAK, P. J. Plasma urate levels in mongolism. *American Journal of Mental Deficiency*, 74(2):196-199, 1969.

This study indicates that the abnormal urate metabolism of the mongoloid previously reported by various authors also involves the

precursors xanthine and hypoxanthine. Significantly higher plasma uric acid, xanthine, and hypoxanthine concentrations have been found in the mongoloid. Possible biochemical relationships of purine biosynthesis, γ -globulin biosynthesis, and vitamin A absorption are also discussed. (25 refs.) - *Journal abstract*.

Department of Chemistry
University of Scranton
Scranton, Pennsylvania 18510

2549 BENSON, PHILIP F., LINACRE, BRIAN, & TAYLOR, ANGELA I. Erythrocyte ATP: D-fructose-6-phosphate 1-phosphotransferase (phosphofructokinase) activity in children with normal/G trisomic mosaic Down's syndrome and in normal and Down's syndrome controls. *Nature*, 220(5173):1235-1236, 1968. (Letter)

Erythrocyte phosphofructokinase activity was significantly correlated with the proportion of trisomic 21 cells in patients with Down's syndrome. Enzyme activity in 10 control patients showed a mean of 0.91 units, and in 10 patients with Down's syndrome the mean was 1.29 units. Six individuals, mosaic for trisomy 21, showed intermediate levels of activity (mean 1.08 units), and this appeared to be in proportion to the degree of mosaicism. (4 refs.) - E. L. Rowan.

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London S. E. 1, England

2550 LAWTON, SHERMAN B., STODDARD, GAYLE R., & SEELY, J. RODMAN. Familial 21/21 translocation. *Journal of Pediatrics*, 74(2):305-309, 1969.

A 21/21 translocation is probable in a family in which 8 consecutive pregnancies produced 3 MR children with Down's syndrome and 5 spontaneous abortions. Peripheral blood samples indicated that the father had a normal 46,XY chromosome complement while the phenotypical mother had a 45,XX,G-G,t(GqGq) karyotype. The 3 children exhibited typical mongoloid stigmas and identical karyotypes--46,XX,G-G,t(GqGq)mat. The paternal and maternal family histories were clear. Although morphological differentiation between the chromosomes in the G-group was not possible, probability evidence [(1/2)⁸] indicates the presence of a 21/21 translocation. (11 refs.) - A. Huffer.

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800 Northeast 13
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- 2551 VETRELLA, M., BARTHELMAI, W., & MATSUDA, H. Down's syndrome and cystic fibrosis. *Pediatrics*, 43(5):905, 1969. (Letter)

A newborn, who died at 49 days of age, had trisomy 21 and cystic fibrosis with hypoplastic thrombocytopenia. The sweat test showed 81 mEq Na/l, 141 mEq Cl/l; there were no pancreatic enzymes in the duodenal juice and meconium passage was delayed. The infant had pneumonia and intestinal problems and, at death, had severe diarrhea, anemia, leukopenia (white blood cells 3,500/mm³, 520 neutrophils), and a platelet count of 2,000/mm³. The combination of Down's syndrome and cystic fibrosis in a S could be a useful model for locating the gene responsible for cystic fibrosis. (3 refs.) - A. Huffer.

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- 2552 Hepatitis and the Australia antigen. *British Medical Journal*, 2(5658):645-646, 1969. (Editorial)

The incidence of Australia antigen in patients with Down's syndrome is much lower if they are seen as out-patients or if they have been cared for in small residential settings. The percentage of patients with Australia antigen and Down's syndrome who can be classified as carriers and those who have chronic inflammation of the liver is unknown. Australia antigen is rarely found in other MR patients in the same institutions, and there are no reported cases on the medical staff. Unanswered is the question of whether persistence of the antigen can cause chronic hepatitis, and further research is needed to develop an effective hepatitis virus vaccine. (12 refs.) - S. Half.

- 2553 MEADOW, S. R., & MacKEITH, R. C. Mongoloid features with normal chromosome-dysplasia epiphysealis punctata. *Proceedings of the Royal Society of Medicine*, 60(9):894-895, 1967.

An infant diagnosed as mongoloid at birth was subsequently found to have normal chromosomes and a radiological picture of dysplasia epiphysealis punctata. She was born after a normal pregnancy and, in addition to multiple mongoloid features, had talipes and a short leg. At 10 months, many mongoloid features remained, including flat occiput, flat face, slanting palpebral fissures, single transverse palmar creases, and short, incurled

little fingers; however, there were abnormalities of the bones not associated with mongolism. The correct diagnosis was made by X-ray of the bones. With time, the mongoloid features have tended to disappear. Since this disease closely resembles mongolism in infancy, it should be kept in mind as a differential diagnosis in newborn infants with mongoloid features. (5 refs.) - W. Klein.

Evelina Children's Hospital of
Guy's Hospital
London, England

- 2554 FRANCOIS, J., MATTON-VAN LEUVEN, M. TH., & COPPIETERS, R. Short arm enlargement in a G chromosome. *Acta Geneticae Medicae et Gemellologiae*, 27(3):468-486, 1968.

Three cases of enlargement in length of the short arm of a G chromosome are reported. A 2-year-old boy with psychomotor retardation, microcephaly, nystagmus, and congenital glaucoma had a karyotype in which there was an odd submetacentric chromosome with a total length approximating that of an F chromosome, a short arm the size of the short arm of chromosome 18, and a long arm the length of a long arm of a G chromosome. This chromosome was assigned to the G group and the identical chromosomal aberration was also found in karyotypes of the 2 1/2-year-old male mongoloid and his normal mother. The odd chromosome can be the result of altered chromosomal behavior with anomalous coiling or a translocation. (22 refs.) - M. G. Conant.

Department of Human Genetics
Ophthalmological Clinic of the University
Ghent, Belgium

- 2555 NUZZO, FIORELLA, MARINI, A., BAGLIONI, C., FORD, C. E., DeCARLI, L., & SERENI, LUCIA PICENI. A case of multiple chromosomal rearrangements with persistence of foetal haemoglobin. *Cytogenetics*, 7(3):169-182, 1968.

Chromosome analysis of mitotic cells from a 15-month-old male infant with MR, psychomotor retardation, and unusual facies revealed 3 abnormal chromosomes. One was identified as a number 3 with an extra segment, one as a number 6 or 7, or X, with a deletion, and the last as a member of pair 15. No evidence of any karyotypic abnormality was found in peripheral blood cells from either parent. Embryonic hemoglobin persisted in the patient at 11 months of age and a fetal hemoglobin level of 26% was found at age 15 months. Postnatal persistence of above normal levels

of fetal hemoglobin has been associated with cases of trisomy 21, and it is possible that one or more of the abnormal chromosomes are carriers of gene(s) controlling the synthesis of ϵ and γ chains. (32 refs.) - M. G. Conant

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Pavia Section
Pavia, Italy

2556 NEU, RICHARD L., & KAJII, TADASHI.

Other autosomal abnormalities. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 9, p. 652-667.

Syndromes associated with partial deletions of autosomes can be roughly divided into 2 groups: those in which there are similar clinical and chromosomal findings, and those in which there are variable clinical features but similar chromosome findings. The first group includes: the short-arm deletion of number 4; the *cri-du-chat* syndrome (short-arm deletion of number 5); the long-arm deletion of number 18; and the long-arm deletion of number 21. The second group includes: the short-arm deletion of number 18; the short-arm deletion of a D-group chromosome; and the short-arm deletion of a G-group chromosome. The most characteristic feature of partial deletion of an autosome is MR. Partial trisomies of 13-15 or 21-22 chromosomes, trisomies of the C-group, trisomy-22, and ring chromosome formation are briefly discussed. Partial deletion of an autosome offers a rare opportunity to locate the genes which control various biological functions (blood groups, haptoglobulins, and Gm factors). (46 refs.) - L. S. Ho.

2557 POLANI, P. E. Autosomal imbalance and its syndromes, excluding Down's.

British Medical Bulletin, 25(1):81-93, 1969.

Half of the abnormalities in surviving infants are caused by autosomal anomalies which can be divided into errors of numbers and errors of structure. Numerical autosome anomalies include Patau's syndrome, Edwards' syndrome, spontaneous abortion, and triploidy, all of which include MR as a clinical feature. Structural autosome anomalies are subdivided into deletions, duplications, and ring chromosomes. Anomalies involving deletions and duplications include deletions of the short

arm of a B-group chromosome (*cri-du-chat* syndrome), deletions of the short arm of an E-group chromosome, deletions of the long arm of an E-group chromosome, simple deletions of a D-group chromosome, and simple deletions of other autosomes, with MR involved in the first 3 subgroups. Anomalies involving ring chromosomes include ring E chromosomes, ring D chromosomes, ring-chromosome formation, deletions or monosomy of a G-group autosome, and other ring autosomes. Other structural autosome anomalies include simple duplications, isochromosomes, and inversions. (228 refs.) - M. G. Conant.

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2558 PUNNETT, HOPE H., & MELLMAN, WILLIAM J.

Familial chromosome translocation. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 9, p. 668-681.

Progeny of a phenotypically normal, translocation-carrier parent may have a balanced translocation, an unbalanced translocation, or a normal chromosome. Translocations are often detected through progeny with multiple anomalies and unbalanced chromosome complements. The typical karyotype pattern contains a heteromorphic chromosome pair; one is normal, the other is too long or too short, or has the centromere in the wrong position. One of the parents will often have this same abnormal chromosome as well as another heteromorphic pair. The classic examples are: the familial occurrence of Down's syndrome involving a D/G translocation and of D-trisomy involving a D/D translocation. The most consistent finding in patients with unbalanced autosomal complements is MR. Microcephaly and cephalofacial disproportion are often accompanying features. The incidence of unbalanced translocation progeny is far lower than expected from the possible chromosomal arrangements at meiosis. This is probably due to the lack of viability of the zygotes of unbalanced translocation and is the probable cause of the high frequency of abortion among pregnancies of balanced carriers. The knowledge of translocation is eugenically significant and should be revealed to the parents. The affected parents may choose to undergo sterilization or, if the male is a carrier, to allow artificial insemination. They may decide to deny the risk and to have more children. Determination of karyotypes of conceptuses by culturing cells obtained by amniocentesis is imperfect at the present time. (30 refs.) - L. S. Ho.

- 2559 FORD, C. E., & CLEGG, HILARY M. Reciprocal translocations. *British Medical Bulletin*, 25(1):110-114, 1969.

Presumptive reciprocal translocations, in which the chromosomes of the propositus and both parents were examined, were ascertained for 129 cases, of which 118 were children with congenital malformations, 6 were women with histories of spontaneous abortion, and 5 were discovered fortuitously. Analysis of the participation of the different chromosome groups showed that break-points are much more frequent than expected in groups B, D, E, and G and less frequent than expected in groups A, C, and F. In the 129 cases, the translocation was inherited from the mother in 65 cases, inherited from the father in 28 cases, the propositus was not a mosaic in 30 cases, and he was a mosaic in 6 cases (parents were karyotypically normal in these 36 cases). The occurrence of normal/translocation mosaics indicates that structural changes can originate in somatic cells during development and are not limited to germ cells. (15 refs.) - M. G. Conant.

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- 2560 HAMERTON, J. L. Robertsonian translocations in man: Evidence for prezygotic selection. *Cytogenetics*, 7(4):260-276, 1968.

Additional data on segregation of the 3 types of Robertsonian translocations, obtained from 33 families with a segregating t(DqGq), 26 families with a segregating t(DqDq), and 4 families with a segregating t(21q22q), are considered. In both the t(DqGq) and t(DqDq) types, there is a significant excess of balanced heterozygotes over chromosomally normal progeny among the offspring of the male heterozygotes. This finding supports the hypothesis that some form of prezygotic selection or meiotic drive in man exists, possibly due to the preferential recovery of certain classes of spermatozoa. In addition, the 2 types of translocations showed a deficiency of unbalanced progeny. In all 3 types of translocation, the frequency of spontaneous abortions is equal to that in the general population. There was no evidence to suggest the association of any particular phenotype with balanced t(DqDq) heterozygotes. (51 refs.) - M. G. Conant.

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- 2561 FORD, C. E. Mosaics and chimaeras. *British Medical Bulletin*, 25(1):104-109, 1969.

Mosaics are formed from the cells of a single zygote lineage and originate as consequences of irregularities during the cell cycle, while chimaeras have cells derived from 2 or more distinct zygote lineages and arise either from 2 separate instances of syngamy in one ovum or from intermixture or exchange of cells between individuals of independent zygotic origin. Chimerism can be proved by genetic evidence, but mosaicism cannot be proved at all, although the cytogenetic evidence is generally striking enough for acceptance. Most reported cases of mosaicism involve the sex-chromosomes, although a few cases of autosome mosaicism have been found. Several examples of undoubted monozygotic twins with markedly different phenotypes have been found. Nine different types of chimaeras are theoretically possible, but only 6 have been identified. Four types involve dispermy and produce distinguishable differences, one type involves fusion of one daughter of zygote nucleus with the nucleus of a second polar body, and 4 types involve contributions from 2 independent embryos (early fusion of 2 embryos, placental cross-circulation between dizygotic twins, maternal-fetal transplacental exchange, and artificial by transplantation or grafting). (36 refs.) - M. G. Conant.

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- 2562 PORTER, IAN H., BROWN, CHARLES D., GERGOSIAN, LORAIN E., & PAUL, BETTY A. Evidence of selection in mosaicism. *Journal of Medical Genetics*, 6(3):310-313, 1969.

A boy who died at 10 months of age exhibited multiple congenital malformations and a selectivity for one-stem-line chromosome constitution at the expense of 4 others. The S was the fifth child of normal Caucasian parents and the product of an uncomplicated pregnancy and delivery. He had cleft lip and palate, dysplastic ears, micrognathia, prominent occiput, coloboma of the left iris, hypotonia, "rockerbottom" feet, 6 fingers on the left hand, right ventricular hypertrophy, umbilical and bilateral inguinal hernias, and bilateral cryptorchidism. Peripheral blood cultures of 93 cells at 2 weeks indicated the presence of 4 stem-lines: 7% were 45,XY,D-,G-,t(DqDq)+; 58% were 46,XY,D-,t(DqGq)+; 30% were 46,XY,D-,E+,G-,t(DqGq)+; and 4% were 47,XY,D-,E+,t(DqGq)+. At 3 months of age, 107 peripheral blood cells showed 3

stem-lines: 22% were 45,XY,D-,G-,t(DqGq)+; 4% were 46,XY; and 92% were 46,XY,D-,t(DqGq)+. Of the 169 blood cells examined at 6 months of age, 8% were 45,XY,D-,G-t(DqGq)+ while 92% were 46,XY,D-,t(DqGq)+. At 8 months, 98% of the 149 blood cells examined were 46,XY,D-,t(DqGq)+. Fibroblasts from skin biopsies taken at 2 weeks and 3 months showed only the 46,XY,D-,t(DqGq)+ chromosome complement. The parents and 4 normal brothers showed no chromosome abnormalities. When the karyotype does not match the phenotype, it is possible that the survival of one stem-line at the expense of another has determined the course of embryonic development. (5 refs.) - A. Huffer.

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2563 MINTZ, BEATRICE. Developmental mechanisms found in allophenic mice with sex chromosomal and pigmentary mosaicism. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part I. Special Lectures.* (Birth Defects Original Article Series, Volume 5, Number 1.) New York, New York, 1969, p. 11-22.

Functional mosaicism and differential gene expression have been studied in the reproductive and pigmentary systems of allophenic mice in which at least 2 cell lines have been "assembled" at the embryo stage. Both XX and XY cells may coexist in the mosaic, but the sex phenotype depends on the genotype composition of gonadal soma, is stable and irreversible, and is independent of mosaicism in other cell systems. Even in the sex glands, a large minority of "wrong sex" cells can be tolerated. The geometric patterns of multicolored allophenic patterns based on clonal migration depend upon clonal selection and interaction. Competition and selection between phenotypically different subpopulations of cells may be a universal device for the determination of the total phenotype. (23 refs.) - E. L. Rawan.

2564 RACE, R. R., & SANGER, RUTH. X and sex chromosome abnormalities. *British Medical Bulletin*, 25(1):99-103, 1969.

The antigen Xg^a is inherited as a sex-linked dominant trait. The distribution of Xg groups in several sex-chromosome aneuploidies are reviewed, including Klinefelter's syndrome (XXY, XXXY, XXXXY, XYY, XXY/males, and XX males), Turner's syndrome, XXX females

XXXX females, and XY females with the testicular feminization syndrome. The observed Xg distribution can be compared with the statistical distribution determined for males and females, and deviations are useful for indicating heterogeneity of background and the site of the accident. The Xg^a, when located on an abnormal X chromosome, is inactivated, and familial studies indicate that the Xg locus must be involved in this inactivation. Study of Xg groups has proved to be very useful in problems of X-chromosome aneuploidy and in structural abnormalities of the X-chromosome. (31 refs.) - M. G. Conant.

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2565 JACOBS, PATRICIA A. Structural abnormalities of the sex chromosomes. *British Medical Bulletin*, 25(1):94-98, 1969.

The present state of knowledge of structural abnormalities of the sex chromosomes is reviewed; the X and Y chromosomes associate terminally at meiosis, but it is not known whether the short arm of the X associates with the long or short arm of the Y. Current evidence suggests that meiosis breaks down when the sex chromosomes fail to associate and that, in persons with an abnormal number of sex chromosomes, a mechanism may exist for the selective loss of the additional chromosome sometime before meiosis, so that only those cells with a normal sex-chromosome complement form gametes. These problems should be elucidated by further study of meiosis in persons with abnormalities of the sex chromosomes. The identification of structurally abnormal sex chromosomes poses some problems, especially for the Y chromosome. Clinical and cytological features associated with structurally abnormal X and Y chromosomes indicate that genes influential in development of stature are situated on the short arm of the X chromosome and that genes responsible for the development of testes are located on the short arm of the Y chromosome. (33 refs.) M. G. Conant.

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Edinburgh, Scotland

- 2566 OHNO, SUSUMU. *Sex Chromosomes and Sex-Linked Genes*. Berlin, Springer-Verlag, 1967, 192 p. \$9.50.

Polyphyletic evolution of vertebrate sex chromosomes, dosage effect of sex-linked genes and effective dosage compensation mechanisms, and the nature of sex-determining factors and the act of sex determination are the topics of this book. Differentiation of an homologous chromosome pair to the sex pair (X and Y in mammals or Z and W in birds) was accomplished by accumulation on one member of the pair most of the factors governing the development of the heterogametic sex. All the Mendelian genes which were originally on it were eliminated by progressive genetic deterioration, and the chromosome eventually emerged as a very specialized sex determiner of minute size. In placental mammals, the X-linked genes exist in the hemizygous state in the male. Each X-linked gene accommodated itself to this state by doubling the rate of product output. Therefore, disparity between the male and the female is very great. The dosage effect of X-linked genes in females was adjusted by randomly inactivating one of the X pair. The sex-determining factors acquired certain peculiarities during differentiation which set them apart from the Mendelian genes. The question of how many kinds of male-determining factors are on the Y chromosome is yet to be answered. These factors express themselves through somatic elements of the embryonic-indifferent gonads. When the male-determining factors prevail, interstitial cells develop and the testis is organized. This book should be of interest to biologists and geneticists. (293 Refs.)
L. S. Ho.

CONTENTS: Evidence Indicating That the X and the Y or the Z and the W Were Originally an Homologous Pair of Ordinary Chromosomes; Differentiation of the Primitive Sex Elements at the Expense of the Y or W, and the Conservation of the Original X or Z; Polyphyletic Evolution of Vertebrates; Conservation of the Original X and Homology of the X-linked Genes in Placental Mammals; Conservation of the Original Z-chromosome by Diverse Avian Species and Homology of the Z-linked Genes; The Basic Difference in Constitution between the Mammalian X and the Drosophila X; The Two Different Means of Achieving Dosage Compensation for X-linked Genes Employed by Drosophila and Mammals; Various Consequences of the Dosage Compensation by X-inactivation; The Conservation of the Original X and Dosage Compensation in the Face of X-polysomy; Three Different Consequences of X-autosome Translocation; The Consequences of Y-autosome Translocation and the XO as the Normal Female of Certain Mammalian Species; Apparent Absence of Dosage Compensation for Z-linked

Genes of Avian Species; Elucidation of So-called Sex-determining Factors; Time and Place of Action of Sex-determining Factors in Ontogeny.

- 2567 JUBERG, RICHARD C., JEWSON, DOUGLAS V., TAYLOR, MARY BELLE, & MOORE, VICKI L. Chromosome studies in patients with hypospadias. *Pediatrics*, 43(4):578-582, 1969.

Twenty-one phenotypic males with hypospadias were subjected to chromosomal analysis, but no abnormalities of number nor structure were demonstrated. This patient group was felt to be a representative sample of cases with hypospadias. Approximately 30 leukocytes were studied in each patient and no evidence of mosaicism was found; however, cultures of the affected tissue were not done. There appears to be no genetic basis for the assumption that hypospadias represents a form of intersexuality. (20 refs.)
E. L. Rowan.

Department of Pediatrics
West Virginia School of Medicine
Morgantown, West Virginia 26506

- 2568 BURCH, P. R. J. Klinefelter's syndrome, dizygotic twinning and diabetes mellitus. *Nature*, 221(5176):175-177, 1969. (Letter)

Dizygotic twinning, diabetes mellitus, and Klinefelter's syndrome appear to have some relation to each other. Data support a connection between Klinefelter's syndrome and dizygotic twinning. Dizygotic twinning does not appear to occur as often after a maternal age of 35; an autoaggressive mechanism may intervene and the same mechanism is cited as probably responsible for the connection between Klinefelter's syndrome and dizygotic twinning. A high prevalence of diabetes mellitus is reported in parents and relatives of patients with Klinefelter's syndrome as well as in the patients themselves. Sex and CA distribution of diabetes mellitus suggest that it is also an autoaggressive disorder. If diabetes mellitus and meiotic non-disjunction of the X chromosome producing Klinefelter's syndrome are genetically-based autoaggressive disorders, then patients with Klinefelter's syndrome should have more cases of diabetes mellitus; this assumption is supported by research. (17 refs.)
B. Bradley.

Department of Medical Physics
University of Leeds
Leeds, England

2569 RICCI, N., DALLAPICCOLA, B., VENTIMIGLIA, BERNADA, TIEPOLO, L., & FRACCARO, M. 48,XXX/49,XXXX mosaic: Asynchronics among the late-replicating X chromosomes. *Cytogenetics*, 7(4):249-259, 1968.

A 13-year-old girl with SMR and multiple malformations was found to be a 48,XXX/49,XXXX mosaic. Oral mucosa smears and cultured fibroblasts revealed multiple sex chromatin bodies (1-4). This is the third reported case of a female having 5 X chromosomes. All 3 had MR, bone deformities, and multiple malformations. There were a low number of cells with drumsticks located in the granulocytes, the incidence being 2/1,000. The bone marrow of this girl also had cells with at least 4 X chromosomes. A low total ridge count was found on the fingertips. Analysis shows an "internal asynchrony among the late replicating X's in about 50% of the cells with 4 & 5 X chromosomes." (11 refs.) - B. Bradley.

Clinica Medica
Universita
44100 Ferrara, Italy

2570 KIHLMOM, MAGNUS. Psychopathology of Turner's syndrome. *Acta Paedopsychiatrica*, 36(3/4): 75-81, 1969.

Eleven cases of Turner's syndrome with chromosome constitution XO have been investigated with child psychiatric methods. Seven were of normal intelligence, 4 were MR. There was no case of psychosis, or epilepsy, or grave symptoms of brain damage. Three cases displayed signs of minimal cerebral dysfunction. All were considered psychoinfantile. The majority had a remarkably low level of aggressiveness and intensity of affect. Neurotic symptoms were present in several cases in conformity with the opinions of Lambert concerning Klinefelter's syndrome and those of Mellbin regarding neuropsychiatric findings in Turner's syndrome, the writer finds good reasons for assuming that the chromosome aberration in Turner's syndrome predisposes for a psychopathology of the aforementioned type. (18 refs.) - *Journal summary*.

Child Psychiatric Clinic of Karolinska
Institutet
Stockholm, Sweden

2571 BUTLER, L. J., CHANTLER, C., FRANCE, N. E., & KEITH, C. G. A liveborn infant with complete triploidy (69,XXX). *Journal of Medical Genetics*, 6(4):413-421, 1969.

A premature female who survived only 23 hours is described. External anomalies included hypertelorism, bilateral microphthalmia, and colobomata, low-set ears, single palmar creases, absent flexion creases of some fingers, and unusual dermatoglyphs. Internal defects included congenital heart disease (atrial and ventricular septal defects, bicuspid aortic and pulmonary valves), cystic hypoplasia of kidney and hydronephrosis, hypoplasia of ovaries, pituitary, and adrenals. Hemolytic disease was possibly due to ABO incompatibility. Chromosome analysis of blood and skin revealed complete triploidy (69,XXX). Studies of nuclear areas confirmed the presence of a single cell population in fibroblast cultures, and only 2.5% of nuclei were observed with 2 sex chromatin bodies. The congenital malformations of other complete and mosaic triploids reported are tabulated and discussed. Reference is made to the imbalance between the sex chromosome complement and the autosomes resulting in intersexuality shown by 69,XXY males. (20 refs.) - *Journal summary*.

Queen Elizabeth Hospital for Children
Hackney Road
London E.2, England

2572 BARR, M. L., SERGOVICH, F. R., CARR, D. H., SHAVER, EVELYN L. The triplo-X female: An appraisal based on a study of 12 cases and a review of the literature. *Canadian Medical Association Journal*, 101(5):247-258, 1969.

An assessment has been made of the effect of the triplo-X (47,XXX and 47,XXX/46,XX mosaic) sex chromosome error on development, based on a study of 12 cases and reports of 143 such females in the literature. It appears that about 1/3 of triplo-X females have congenital defects of a physical nature. The majority are physically normal, and because the defects vary so greatly in type and severity, there is no triplo-X syndrome in the clinical sense. Although the chromosome error predisposes to ovarian dysfunction or pathology to some extent, the majority of triplo-X females have a normal reproductive system and 28 of them have borne 67 children.

Mental aspects of the triplo-X phenotype are especially difficult to assess because of the methods of ascertainment. Most of the patients affected were found in the course of surveys of populations of MR or mentally ill females. The risk of MR or mental illness is undoubtedly high, although a proportion of triplo-X females, the magnitude of which is at present unknown, are normal mentally. The psychopathology associated with this chromosome abnormality is especially in need of further study. Trisomy of the X chromosome has in general a much less adverse effect on development than trisomy of any autosome. This is attributed to heteropycnosis and functional suppression of X chromosomes in excess of one. (141 refs.) - *Journal summary*.

University of Western Ontario,
London, Ontario
Canada

- 2573 CHRISTAKOS, ARTHUR C., SIMPSON, JOE LEIGH, YOUNGER, J. B., & CHRISTIAN, C. D. Gonadal dysgenesis as an autosomal recessive condition. *American Journal of Obstetrics and Gynecology*, 104(7):1027-1030, 1969.

The occurrence of gonadal dysgenesis and a normal female (46/XX) chromosomal complement in 3 MR sisters whose parents are consanguineous is probably due to autosomal recessive inheritance. The Ss were slightly under 5 feet in height, had no somatic abnormalities of Turner's syndrome, had not developed secondary sexual characteristics, have had no menarche, exhibited retarded bone age, had elevated urinary gonadotropins, showed no abnormalities in urinary 17-hydroxy and 17-ketosteroids, possessed normal intravenous pyelograms, had normal skull films, exhibited normal fields of vision, showed no mosaicism in lymphocytes or fibroblasts, and revealed streak gonads. In addition, a deceased brother and 2 of the patients had bilateral sensory hearing loss. (29 refs.) - A. Huffer

Department of Obstetrics and Gynecology
Duke University School of Medicine
Durham, North Carolina 27706

2574 MOOR, LISE. Niveau intellectuel et polygonosomie: Confrontation du caryotype et du niveau mental de 374 malades dont le caryotype comporte un excès de chromosomes X ou Y (The intelligence level and polygonosomia: A comparative study of the karyotype and the mental level in 374 patients with an excess of X or Y chromosomes in the karyotype). *Revue de Neuropsychiatrie Infantile et d'Hygiène Mentale de l'Enfance*, 15(4-5):325-348, 1967.

A review of the literature on studies of 64,335 Ss showed that polygonosomia is 4-5 times more frequent among the MR than among the general population, 0.39% versus 0.085% for females and 0.89% versus 0.22% for males. Also, the frequency of polygonosomia is twice as high among the mildly and moderately retarded as among the severely retarded. The average IQs of 374 patients with polygonosomia ranged from 35.2 to 83.9, with MR becoming more severe as the number of gonosomes in the karyotype increased and as the number of X chromosomes in excess increased. Although a mathematical correlation between karyotype and IQ cannot be established at the present time, it appears MR is most profound in cases with the most X chromosomes and, with an equal number of X chromosomes, is more severe with increasing number of Y chromosomes. (158 refs.) - M. G. Conant.

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47, bd de l'Hopital
Paris 13^e, France

- 2575 CLOSE, H. G., GOONETILLEKE, A. S. R., JACOBS, PATRICIA A., & PRICE, W. H. The incidence of sex chromosomal abnormalities in mentally subnormal males. *Cytogenetics*, 7(4): 277-285, 1968.

In MR males, an abnormal sex chromosome complement may be related to behavioral disturbances and above average height. Of 917 men examined, 19 were 6 feet or more in height and 5 of these had abnormal sex chromosomes. Forty-nine were between 5 feet 10 inches and 6 feet in height; 2 of these were found to have abnormal sex chromosomes. Eleven of the total 917 (1.2%) were chromatin positive. Four of 82 men (4.7%) detained under court order were chromatin positive compared to 7 of 824 (0.9%) informally admitted. (15 refs.) - L. S. Ho.

*Western General Hospital
Edinburgh 4, Scotland

2576 SPENCER, D. A. An unusual sex chromosome mosaicism. *British Journal of Psychiatry*, 115(523):747, 1969. (Letter).

A 22-year-old male MR, who presented the mental and physical features of Klinefelter's syndrome and the XYY syndrome, had cell lines with chromosomal complements of 47XYY, 48XXYY and 49XXYY (in one cell). The S is 73 inches tall, weighs 132 pounds, has a normal EEG, and has a history of aberrant behavior. His excretion of 17-ketosteroids and 17-hydroxycorticosteroids is below normal. His fingerprints showed ulnar loops except for whorls on both ring fingers and the right index and middle fingers. (No refs.) - A. Huffer.

Westwood Hospital
Cooper Lane
Bradford, 6 England

2577 MOORE, G. E., PORTER, I. H., & HUANG, C. C. Lymphocytoid lines from persons with sex chromosome anomalies. *Science*, 163(3874): 1453-1454, 1969.

To establish permanent lymphocytoid cell lines, leukapheresis was performed on blood from a 28-year-old MR male (who exhibited antisocial behavior and had a history of suspected arson) with an XYY phenotype and a 31-year-old male with Klinefelter's syndrome. Respective distribution of chromosome counts of 45, 46, and 47 were: 0, 9, 92 for one cell line from the 46 XYY constitution; 1, 1, 48 for the other cell line of the same patient; and 2, 18, 80 for the S with Klinefelter's syndrome. A library of such cell lines and cell lines from individuals with other genetic defects will provide important viral, biochemical, immunological, and clinical information. (9 refs.) - A. Huffer.

Memorial Institute
New York State Department of Health
Buffalo, New York 14203

2578 NIELSEN, J., YDE, H., & JOHANSEN, K. Serum growth hormone level after oral glucose load, urinary excretion of pituitary gonadotrophin and 17-ketosteroids in XYY syndrome. *Metabolism: Clinical and Experimental*, 18(12):993-997, 1969.

No significant indications of growth hormone overproduction for the fasting state or after oral glucose load were found for 6 patients with the XYY syndrome. Fasting serum growth hormone, serum growth hormone response to an oral glucose load, and urinary excretion of

pituitary gonadotrophin, 17-ketosteroids, and fractionated 17-ketosteroids were measured for 6 patients. Five patients were in psychiatric institutions with an additional case in an institution for the MR. The CAs ranged from 16-39 with a mean of 26 years. Mean height was 188 cm and extended from 182 to 200 cm. Control group included 13 non-obese persons with a mean CR of 36 years and a range from 19-54 years. Results showed no difference in mean fasting serum growth hormones for the 2 groups. Four of 5 patients with the XYY syndrome were within the normal range and urinary excretion of pituitary gonadotrophin was in the normal range in 3 of the 6 patients. (18 refs.) - B. Bradley.

Cytogenetic Laboratory
Aarhus State Hospital
Risskov, Denmark

2579 STENCHEVER, MORTON A., & MACINTYRE, M. NEIL. A normal XYY man. *Lancet*, 1(7596):680, 1969. (Letter)

A 6-foot-tall Caucasian male (CA 34 yrs) who had no criminal record and was described as a mild-mannered, hard-working, responsible, high school graduate has a 47,XYY karyotype. Since the XYY syndrome has legal implications, all normal Ss with such a chromosome constitution should be documented. (1 ref.)
A. Huffer.

Case-Western Reserve University
Cleveland, Ohio 44106

2580 COWLING, D. C., RIGO, S., & MARTIN, F. I. R. The "YY syndrome," *Medical Journal of Australia*, 2(10):443-446, 1969.

A 20-year-old male (IQ 95) who had an XYY chromosomal constitution was 6 feet 5 1/2 inches tall and had severe varicose veins; however, he had no antisocial behavior. Although he was somewhat obese, physical examination revealed no unusual findings; specifically, his body hair distribution, testes, and breasts were normal. Blood pressure and pulse, electrocardiogram, X-ray of the pituitary fossa, glucose tolerance curve, and urinary excretion of pituitary gonadotrophins were normal. Illness caused him to terminate each of the 4 jobs he had had after receiving

his technical school certificate, although he held the last job 4 years. Supportive psychotherapy and anti-anxiety agents were required as a result of the stripping operation for the varicose veins. (20 refs.)

A. Huffer.

Royal Melbourne Hospital
Melbourne, Victoria 3050,
Australia

- 2581 SUINN, RICHARD M. YY syndrome and sampling techniques. *Lancet*, 1(7586):157-158, 1969. (Letter)

Since Ss in previous reports were usually selected on the basis of being institutionalized and tall when the relation between the YY syndrome and criminality was investigated, a different sampling technique based on the presence of the XYY condition alone is now appropriate. This sample would test the relation between the syndrome and criminal behavior by examining for false positives. If there is an invariable relation, there should be no false positives; however, the finding of a false positive would not rule out an association, but, the possession of such a karyotype would carry less weight in a court trial. (8 refs.) - A. Huffer.

Department of Psychology
Colorado State University
Fort Collins, Colorado 80521

- 2582 MORRIS, T. The XO and OY chromosome constitutions in the mouse. *Genetical Research*, 12(1):125-137, 1968.

The mouse is an excellent subject in which to study sex chromosome abnormalities because even though the OY constitution is lethal, the XO mouse is viable, fertile, and phenotypically normal. Litters of XO and XX females were compared at weaning, birth, and 15 and 3 1/2 days gestation. Evidence suggests that all OY zygotes are lost before implantation and can be seen as abnormal (arrested development at the 2-cell stage) zygotes only when the uterus is washed 3 1/2 days after coitus. XO zygotes are lost at all stages of embryonic development and observed as small moles (4-9 days post-implantation) and large moles (9-12 days post-implantation) at 15 days in addition to the preimplantation loss. There does not appear to be an excess XO mortality after 12 days gestation. The XO

female (mated to a normal XY male) loses half of all male embryos and 2/3 of all female embryos and the reduced litter is evenly divided among XO, XX, and XY offspring. (16 refs.) - E. L. Rowan.

Medical Research Council
Radiobiological Research Unit
Harwell, Didcot, England

- 2583 NEU, RICHARD L., & GARDNER, LYTT I. Abnormalities of the sex chromosomes. In: Gardner, Lytt I. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 9, p. 682-703.

There is an increased frequency of MR found in patients with Turner's syndrome, Klinefelter's syndrome, poly-X females, and poly-Y males when compared to the normal population. Gonadal dysgenesis, short stature, and sexual infantilism are basic features of Turner's syndrome, while sterility and gynecomastia are common among males with Klinefelter's syndrome. Isochromosome Y, pericentric inversion of Y, and partial deletion of Y have also been reported. There is an increasing proportion of XO cells with age in both males and females; about 7% of XO cells are found in blood cultures of females after the age of 55, and 1-2% in males after 65. (101 refs.) L. S. Ho.

- 2584 LUBS, H. A. A marker X chromosome. *American Journal of Human Genetics*, 21(3):231-244, 1969.

A study of 3 generations of one family revealed the presence of a marker sex chromosome which was associated with MR when it appeared in male offspring. Originally a 2-year-old boy was studied because of MR and multiple minor anomalies. A previously undescribed chromosomal variation consisting of constrictions near the end of the long arms of a C-group chromosome was noted. Subsequently, a family study involving 12 members found 4 male members with the chromosomal variation and borderline to severe retardation (only the original case had anomalies). Three female members of the family showed the chromosomal variation, but were both physically and mentally normal. Labeling studies, arm length studies (measurements), and pattern of appearance of the MR pointed to the X chromosome as being involved. The finding of a marker permitted the identification of the X chromosome for the first time. The detection

of this marker chromosome *in utero* in this family line would permit identification of future males with a high probability of MR. (14 refs.) - W. J. Klein.

Departments of Medicine and Pediatrics
Yale University School of Medicine
New Haven, Connecticut 06510

2585 BENIRSCHKE, KURT. Models for cytogenetics and embryology. *Federation Proceedings*, 28(1):170-178, 1969.

Animal models for cytogenetic and embryologic studies are reviewed with respect to abortion, chimerism, translocation and hybridization, sex chromosomes, embryogenesis, teratogenesis, twinning, and sex differentiation. Examples of suitable models were given for: abortion (pigs); chimerism (cats); translocation (horses including a range from *Equus przewalskii* to *E. zebra hartmannae*); hybrid sterility (mules and male cattle X bison crosses); sex chromosome evolution and cytologic mechanism of dosage compensation (North American flying squirrels); teratogenesis (golden hamsters); twinning (9-banded armadillo); and sex differentiation (rabbits, rats, and chicks). (76 refs.) - L. S. Ho.

Department of Pathology
Dartmouth Medical School
Hanover, New Hampshire 03755

2586 MELLMAN, WILLIAM J. The genetic basis for the variability of hereditary diseases. *Journal of Pediatrics*, 72(5):727-736, 1968.

Evidence of genetic heterogeneity (variable causation) has resulted in the subclassification of 4 clinical problems previously thought to be single genetic entities. Congenital nonspherocytic hemolytic anemia may result from mutations of at least 7 different enzymes concerned with red cell glycolysis and from multiple alterations in the structure of glucose-6-phosphate dehydrogenase. Cystinuria has been divided into 3 types on the basis of variable dibasic amino acid excretion in heterozygotes and intestinal transport in homozygotes. Mucopolysaccharidoses are of 6 types which vary in clinical manifestations, pedigree pattern, and the chemical nature of the urinary metabolite. The oral-facial-digital syndrome is really 2 clinical types with significant pedigree differences--one is transmitted as a dominant lethal to males and the other as an autosomal

recessive. Knowledge about heterozygous patterns is essential in determining prognosis, treatment, and genetic counseling. (27 refs.) - E. L. Rowan.

Department of Pediatrics
University of Pennsylvania
Philadelphia, Pennsylvania 19104

2587 NEU, RICHARD L. Cytogenetic techniques and terminology. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 9, p. 593-607.

Terms used in human genetics such as karyotype, nondisjunction, trisomy, monosomy, translocation, and chromatin are defined, and techniques for studying chromosomes which include preparation of buccal smears, peripheral blood leukocyte culture, bone marrow cell preparation, and skin culture are described in detail. Autoradiographic techniques for studying replication of DNA are also described. The standard nomenclature in human cytogenetics proposed at the Third International Congress of Human Genetics at the University of Chicago are listed. (36 refs.) - L. S. Ho.

2588 New aspects of human genetics (symposium). *British Medical Bulletin*, 25(1): 1-114, 1969. (Available from British Council, London, England) \$6.50.

The classical method of studying human genetics, which is to search for Mendelian inheritance patterns, has been applied to porphyria and to homocystinuria, while the polygenic approach has been successfully applied to the study of certain congenital malformations (pyloric stenosis and anencephaly) and polymorphisms. The polygenic approach stresses the continuously distributed genetic predisposition beyond which individuals are at risk and has proved very useful in the study of familial conditions. The chemistry of the A, B, and H red blood cell antigens has suggested that each person has a unique enzyme constitution, and it has also been found that different populations show hemoglobin variants. The great variety of autosomal abnormalities is reviewed, and new advances in the study of human chromosomes, including balanced reciprocal translocation and the widespread occurrence of chromosome aberrations that are often lethal, are discussed. Human cytogenetics is a relatively new field in which the karyotype abnormalities do not produce Mendelian ratios in families, and 2

studies, concerning the fact that the Xg locus lies on the X chromosome and the linkage of genes on the autosomes, are reported. Current research in the field of human genetics is varied and fruitful, and there are countless areas still open to inquiry. (848 refs.) - M. G. Conant.

CONTENTS: Enzyme and Protein Polymorphism (Penrose); Variations in Structure of Human Haemoglobin (Harris); Genetics of the XX Thalassemias (Lehmann & Carrell); Aspects of Human Blood-group Specificity (Morgan & Walkins); Inborn Errors of Amino-acid Metabolism (Scriver); Homocystinuria (Cusworth & Dent); The Porphyrias (Deen); Genetics of Common Disorders (Carter); Familial Predisposition in Man (Edwards); Progress in Mapping Human Autosomes (Renwick); Human Population Cytogenetics (Court-Brown & Smith); Autosomal Imbalance and Its Syndromes, Excluding Down's (Polani); Structural Abnormalities of the Sex Chromosomes (Jacobs); Xg and Sex Chromosome Abnormalities (Race & Sanger); Mosaics and Chimaeras (Ford); Reciprocal Translocations (Ford & Clegg).

2589 EDWARDS, J. H. Familial predisposition in man. *British Medical Bulletin*, 25(1):58-64, 1969.

Genetic models for conditions not explicable in simple genetic terms include the single-factor theory, which assumes the arbitrary allocation of a constant penetrance unaffected by the genetic background, and the many-factor approach, with reality lying somewhere in between. The technical controversy between the proponents of these 2 extreme approximations cannot be solved by the accumulation of familial data, but requires a knowledge of the mechanism involved. Methods which give a useful summarizing index by analysis of population incidence and incidence in first-degree relatives of affected persons are discussed and include the analytical methods devised by Pearson, Everitt, Falconer, and Edwards. The rough approximations obtained with these methods relate to a disease within the environment studied and give results of limited value. Environmental modification is difficult to evaluate. (25 refs.) - M. G. Conant.

Department of Social Medicine
University of Birmingham
Birmingham, England

2590 CARTER, C. O. Genetics of common disorders. *British Medical Bulletin*, 25(1):52-57, 1969.

The genetic element in most common disorders is not chromosome abnormalities or mutant genes, but probably is an underlying polygenically determined and continuously distributed genetic predisposition with a threshold beyond which individuals are "at risk." Some congenital malformations that appear to be determined in this way are cleft lip (with or without cleft palate), pyloric stenosis, talipes equinovarus, congenital hip dislocation, spina bifida cystica, anencephaly, and congenital heart defects. Family patterns in the incidence of major psychoses (schizophrenia and manic-depressive psychosis), early-onset ischemic heart disease, rheumatoid arthritis, ankylosing spondylitis, and diabetes mellitus suggest that these diseases are also polygenically determined. In polygenic inheritance, the risk to relatives varies from family to family, the more severe degrees of malformation carry a higher risk to relatives, there is an increase in parental consanguinity, and the risk in relatives is absolutely greater than in the general population, but proportionately less. (63 refs.) - M. G. Conant.

Clinical Genetics Research Unit
Hospital for Sick Children
London, England

2591 COURT-BROWN, W. M., & SMITH, P. G. Human population cytogenetics. *British Medical Bulletin*, 25(1):74-80, 1969.

Methods of obtaining more information on human population cytogenetics, which is concerned with the frequency of chromosome aberrations, their causation, and the selective forces that may be exerted on their carriers, were discussed. Retrospective studies involve the comparison with normals of such data as birth order, maternal age, exposure to environmental agents, and infection in the karyotypically abnormal child. Incidence studies are based on the premise that if the survey of some specialized group reveals an aberration frequency in excess of that found in the newborn, a special risk is associated with this aberration. Prospective studies in which individuals with abnormal karyotypes are identified, matched with controls, and subjected to long-term follow-up, have great potential benefit in terms of predictability. The main problems involved in the cytological techniques are the efficiency of detection of structural heterozygotes and difficulties in nuclear sexing. Chromosome studies can also

be used to determine whether a population has been exposed to a chromosome-damaging agent. (35 refs.) - M. G. Conant.

Western General Hospital
Edinburgh, Scotland

2592 CHILDS, BARTON, & DER KALOUSTIAN, VAZKEN M. Genetic heterogeneity. *New England Journal of Medicine*, 279(22):1205-1212, 1968.

The trend is toward a new awareness of the extent of human genetic variation and the recognition that a given character can be due to one of many alleles. Thirty-eight protein types, of which more than one genetically determined variety are known, have been identified and each variant behaves as a Mendelian character, thus inferring the existence of an allelic gene. Genetic tests for allelism in humans usually depend on the fortuitous discovery of families in which 2 variants are found to segregate. More than 150 hemoglobin variants have been discovered; these provide useful insights in visualizing variation at other genetic loci. Of all these variants, only 4 are common in any population. It would be helpful to know the extent of allelic variation in individuals and in populations in order to formulate ideas regarding selection and other factors which determine gene frequency. (96 refs.) - M. G. Conant.

Department of Pediatrics
Johns Hopkins University School of Medicine
Baltimore, Maryland 21205

2593 GALPERIN, HELENE. Comparative study of the association of human acrocentric chromosomes in male and female mitoses. *Cytogenetics*, 7(6):447-454, 1968.

The square distance between the centromeres of the 46 chromosomes was determined on 50 normal metaphase plates, 33 of which were obtained from 8 adult men and 17 of which were obtained from 5 adult women. The square distance values between the chromosomes of group D and group G are significantly smaller than the square distance values taken 2 by 2 from among the 46 chromosomes, and this is much more apparent in females than in males. In addition, the smaller square distance values between chromosomes of group D and those of group G and between all chromosomes of groups D and G are significantly more frequent in

women than in men. This may be an explanation for the important part played by the mother in mongolism which has been attributed to nondisjunction or translocation of chromosome 21. (10 refs.) - M. G. Conant.

Universite Libre de Bruxelles
97 Rue aux Laines
Brussels 1, Belgium

2594 CULBERTSON, JOE C., BRESLAU, NEIL A., MOORE, M. KENT, & ENGEL, ERIC. Sex chromatin determination from hair. *Journal of the American Medical Association*, 207(3):560-561, 1969. (Letter)

Sex-chromatin patterns in cell nuclei can be clearly shown microscopically by aceto-orcein staining of the root sheath from freshly pulled capital hair. Control Ss (10 males; 10 females) had a sex chromatin positivity of 65.1% in females and 1.9% in males. In these controls, and all but one of 330 MR Ss (30 with Down's syndrome), sex chromatin patterns matched phenotypic sex. Deviations from the normal pattern (Klinefelter's syndrome, pseudohermaphroditism, gonadal dysgenesis) were confirmed by clinical and fibroblast studies. (1 ref.) - E. F. MacGregor.

Vanderbilt School of Medicine
Nashville, Tennessee 37205

2595 VIG, B. K., KONTRAS, STELLA B., & PADDOCK, E. F. 3H-thymidine-induced chromosome aberrations in cultured human leukocytes. *Cytogenetics*, 7(3):189-195, 1968.

One-hundred human leukocyte cells from each of 2 patients were treated with tritiated-thymidine, and the incidence of induced chromosomal aberrations was compared with that in 100 untreated cells from each patient. Patient 1 was a 13-month-old male whose karyotype revealed partial deletion of the short arm of a B-group chromosome, and Patient 2 was a 1-month old male with a Philadelphia chromosome in some cells accompanied by anti-mongoloid symptoms. The frequency of aberrations/cell was 0.08 in Patient 1, compared with 1.9 in Patient 2. Using autoradiographic techniques, it was found that aberrations arise with equal frequency in both the synthesizing regions of chromosomes and the regions which have completed DNA syntheses. Gaps attributed to subchromatid breaks are randomly located, not necessarily restricted to late-replicating, heterochromatic regions. Only one transformation, in Patient 2, was

observed. The different responses by the patients to tritiated-thymidine was attributed to differences in genetic background. (14 refs.) - M. G. Conant.

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2596 Glue sniffing may alter chromosomes.
Journal of the American Medical Association, 207(8):1441-1442, 1448, 1969.

Sniffing of toxic agents including airplane glue, nail-polish remover, paint thinner, lighter fluid, gasoline, anesthetics, and nitrous oxide can cause permanent chromosome damage as well as temporary liver damage. A chromosome abnormality rate of 6% was found in 780 cells from 14 patients who were glue or solvent sniffers while there was no chromosome damage in a control group. Of 30 patients, 7 had definite abnormal alkaline phosphatase values and 6 were possibly abnormal. Of 11 patients, 10 showed some liver abnormality on light microscopy, and of these, 7 had abnormal liver function tests. Although 17 of 30 showed minor liver injury, complete recovery was expected if sniffing stopped. Only one patient in 24 had a good relation

with his father, 54.4% lived with only one parent as against 20% in the control group. (No refs.) - M. Plessinger.

2597 CARAKUSHANSKY, GERSON, NEU, RICHARD L., & GARDNER, LYTT I. Lysergide and cannabis as possible teratogens in man. *Lancet*, 1(7586):150-151, 1969. (Letter)

An infant, whose mother was believed to have been exposed to lysergide and cannabis during pregnancy, had terminal transverse deficits of the fingers on the left hand and syndactyly and shortened fingers on the right. X-rays revealed 3 phalangeal bones missing in the left hand and 5 missing in the right hand. The right foot showed webbing between the second and third toes, and the left foot had talipes equinovarus. Chromosomal studies of 31 metaphases revealed no broken chromosomes; 30 cells were 46,XX and one was 45,XX,G-. (3 refs.) - A. Huffer.

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MEDICAL ASPECTS - MISCELLANY

2598 Teeth may testify to retardation. *Medical World News*, 10(16):32, 1969.

By studying tooth development, etiology, hidden CNS damage and time of onset of the insult are possible applications of the relation between dental defects and MR. In one institution, crown defects were found in 74% of the mongoloids, 46% of the brain

damaged, 54% of the cultural-familial MR, and 8% of a normal control group. The number of tooth defects/child was 1.5% for control Ss, 15% for mongoloids, 5% for brain damaged, and 7% in cultural-familial MRs. Certain defects correlate with the type of MR--mongoloids had more canine defects while other MRs had more molar abnormalities. (No refs.) - M. Plessinger.

2599 CLIFF, MAY M., LAPAYONKER, MARC S., & WOLOSIN, HENRY J. Congenital abnormalities of the temporal bone. *Seminars in Roentgenology*, 4(2):122-128, 1969.

Since speech development usually begins before 2 years of age, it is vitally important for the S to be able to hear at that time. Surgical, mechanical, and training programs for the deaf should be instituted. Deafness is often found in the syndromes of Treacher-Collins, Hurler's, osteogenesis imperfecta, Goldenhar, Crouzon, hemifacial microsomia, Waardenberg, and congenital rubella. The Frey and Valvassori classifications of temporal bone abnormalities as well as radiologic determinations of congenital bone abnormalities are discussed. (14 refs.)

R. K. Butler.

Temple University Hospital
Philadelphia, Pennsylvania 19140

2600 SIDMAN, RICHARD L. Abnormal cell migration in developing brains of mutant mice. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 6, p. 40-49.

Studies of the cell migration of recessive mutant "reeler" mice indicate that these animals have abnormal cortical cells which arise at the normal embryonic stages. Autoradiograms of adult mutant and control brains demonstrate that although cortical cells arise over the normal time span in the reeler, they do not assume their normal positions or follow an "inside-out" sequence; cells arising in a given day in the reeler become distributed through all the cortical layers. Studies of embryos killed at different times after injection of thymidine reveal that mutants could be distinguished from normals by the distribution of unlabeled neurons in the developing cortex as early as embryonic day 15. Neurons in the reeler arise in the correct place and undergo normal migration for approximately 48 hours; however, by 72 hours, they have assumed distinctly abnormal positions in the developing cortex. Abnormal function is presumed to be the result of disordered neural circuitry and not of abnormal cell migration *per se*. The reeler provides a relevant model for human diseases which feature cortical dysgenesis in that it is a genetically controlled disorder of neuron migration which results in abnormal circuitry and deranged neurological function. (2 refs.) - J. K. Wyatt.

2601 GLUECKSOHN-WAELSCH, SALOME. Genetic control of neurogenic mechanisms. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 7, p. 50-54.

Mechanisms of neurogenesis which operate at different levels of development and are genetically controlled include the induction of the nervous system during the very earliest embryonic stage, the formation of the neural folds and their rolling into a neural tube, the sorting of neural elements from those of the glia, and the differentiation of embryonic neuroblasts into functional neurons. When any part of the system of regulatory forces needed for the proper operation of neurogenic mechanisms during development is missing or deficient, structural and/or functional abnormalities may result. Human and animal studies indicate that gene mutations can interfere with differentiation of the nervous system at any level of neurogenesis. They can cause primary abnormality of the notochord mesoderm or abnormal cytoplasmic bodies in the neuronal cytoplasm, and they can interfere with the closure of the neural folds into a tube or with normal maturation of the nerve cell. In addition to gene-controlled abnormal differentiation and maturation of the nervous system which are the result of inherent abnormalities either in the differentiating cells and tissues or in their interaction, a large variety of genetic factors affect the nervous system by way of generalized metabolic disturbances. These can exert noxious effects at the level of maturation of inherently normal nerve cells; phenylketonuria and galactosemia are but 2 examples of this model. (No refs.)

J. K. Wyatt.

2602 ALTMAN, JOSEPH. The postnatal origin of microneurons with some evidence of their selective susceptibility to harmful environmental influences. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, Chapter 17, p. 134-136.

Studies of rats established that the bulk of the microneurons which form discrete granular layers in various brain regions are formed after birth. Data from animals with short survival periods after injection with tritiated thymidine revealed a considerable rate of cell multiplication in neonate and infant rats, and a lower, but significant, rate of multiplication in adults. Autoradiographic studies of animals with prolonged post-injection survival periods revealed that cells

which multiplied in the germinal wall of the olfactory ventricle at a high rate in neonate and infant rats migrated into the granular and mitral layers of the olfactory bulb at a speed of approximately 50m/day, and then appeared to become differentiated into granular nerve cells. The germinal wall of the lateral ventricle and the basal polymorph cell layers of the dentate gyrus of the hippocampus also evidence a high rate of cell multiplication in neonate and infant rats. Most of the microneurons in the molecular and granular layers in the rat are of postnatal origin. The postnatal origin of the granule cells indicates that these inter-neurons may represent the plastic elements of the mammalian nervous system and that the synaptic connections of its elements may depend on environmental influences during postnatal migrations and differentiation. Immature cells in the process of differentiation and migration appear to be more susceptible to certain noxious chemical and physical agents than mature, differentiated cells. (3 refs.)

J. K. Wyatt.

2603 SCHMITT, FRANCIS O. The dynamic role of proteins in neuronal membrane functions. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A symposium*. Springfield, Illinois, Charles C. Thomas, Chapter 18, 1968, p. 137-139.

Among the vital functions of neurons and their nets which are subserved by proteins is the process by which impulses are propagated over nerve fibers from the cell body terminals at the synaptic junction, the neuronal membrane systems at the terminals of nerve fibers, and the dendritic and somatic postsynaptic mechanisms. It is hypothesized that, when the protein of the film monolayer undergoes fast conformational change, the lipid bilayer undergoes a phase change and produces local lipid film compactions and rarefactions. The formation of neuronal nets, by aggregation of individual neurons at synaptic junctions, may be due to molecular recognition protein. When the principle of electrogenic protein is applied to the postsynaptic mechanism, it is suggested that the type of potentials recorded by single cell methods may be due to molecular changes in the inner side of the neural membrane. There may be a close coupling between the active biosynthetic processes of the neuron cell body with the membrane and its bioelectric manifestations. (No refs.)

J. K. Wyatt.

2604 EISEN, A. A., & NORRIS, J. W. Adrenal steroid therapy in neurological disease: Part I. *Canadian Medical Association Journal*, 100(1):27-30, 1969.

Adrenal steroids and the adrenocorticotrophic hormone (ACTH) are widely used in a variety of diseases with neurological and psychiatric manifestations. Although their effect is non-specific, success is usually attributed to the "anti-inflammatory" properties of these drugs. Collagen diseases (systemic lupus erythematosus, rheumatoid arthritis, polyarteritis nodosa, scleroderma, polymyositis, temporal arteritis, and thrombotic thrombocytopenic purpura) all respond in some degree to steroid therapy. Known steroid-induced myopathy suggests that these medications are contraindicated in muscle diseases; however, withdrawal after high doses of ACTH has been of some value to patients with severe myasthenia gravis. ACTH in substantial dosage is also useful for acute exacerbations of multiple sclerosis (particularly in reticulobulbar neuritis). (47 refs.) - E. L. Rowan.

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Montreal 2, Quebec

2605 MILLER, ROBERT W., & TODARO, GEORGE J. Viral transformation of cells from persons at high risk of cancer. *Lancet*, 1(7585): 81-82, 1969.

It is now well established that tissue cultures of skin fibroblasts from individuals with a high risk of cancer are especially sensitive to transformation by oncogenic viruses. Fibroblasts from patients with Down's syndrome (G-trisomy) and Fanconi's aplastic anemia, both of which predispose to leukemia, are 3 to 50 times more sensitive than normal fibroblasts to transformation by the SV40 virus. In Fanconi's anemia the range of susceptibility is similar for homozygotes and heterozygotes. The relation between cancer susceptibility and tissue transformation must be studied in such abnormalities as chromosomal disorders (Bloom's syndrome, autosomal trisomies, sex chromosome aberrations, mosaicism, balanced and unbalanced translocations, and DNA damage) inborn immunological defects, congenital growth excesses, and environmental exposure to such oncogenic agents as ionizing radiation and benzene. If these subpopulations at high risk of transformation are affected by environmental factors such as viruses, radiation, or chemical carcinogens, then preventive measures must be taken. (25 refs.) - E. L. Rowan.

National Cancer Institute
Bethesda, Maryland 20014

2606 GRABOW, JACK D., CHUN, RAYMOND W. M., MATTHEWS, CHARLES G., & THOMPSON, WAYNE H. Evolution of cortical electroencephalographic abnormalities to the secondary bisynchrony pattern. *Practical Journal of Psychiatry and Neurology*, 30(7):460-463, 1969.

Evolution of focal cortical spike discharge into a secondary bisynchrony pattern was noted in a S with California encephalitis. Serial EEG tracings were performed during a 22-day hospitalization period in which the S progressed from episodes of psychomotor convulsions and confusion to hemiparalysis, and eventually to relative recovery. Intelligence tests performed before hospital release showed impaired psychometric levels and reduced cognitive functions. A full-scale Wechsler Intelligence Scale for Children (WISC) IQ of 75 was obtained, and the verbal IQ was higher than the performance IQ. Serial EEG tracing at an 8-month follow-up showed spike, sharp wave, and wave activity in the right frontal region. In addition, even bisynchronous bifrontal spike discharges were observed. After 17 months, the WISC IQ was 79; however, overall performance was unchanged. In a follow-up, 2 years later, the EEG tracing showed patterns of secondary bisynchrony. Epileptogenic abnormalities appeared to spread from the right frontal to the left frontal areas in this patient, this suggests that bilateral cortical involvement might be necessary for the development of secondary bisynchrony. Similar findings have been noted in 2 other cases of California encephalitis. (7 refs.) - R. K. Butler.

University of Wisconsin Medical Center
Madison, Wisconsin 53706

2607 PRIBYLOVA, H., & ZNAMENACEK, K. Oxygen consumption and other regulative mechanisms of energy metabolism in pathological states of the newborn. *Biologia Neonatorum*, 14(3-4):133-141, 1969.

In several groups of healthy and pathological newborn infants, oxygen consumption, carbon dioxide output, the respiration quotient, blood levels of glucose, of lactate and pyruvate have been followed-up. Oxygen consumption in premature hypotrophic newborns and in newborns having suffered from intrauterine fetal distress was lower than in healthy ones. Increase in oxygen consumption

in cold which is usually found in healthy newborns was also noted in the majority of newborns having suffered from intrauterine hypoxia. At the same time the glucose level and excess lactate in blood rose in a cold atmosphere while the respiration quotient dropped. Factors influencing oxygen consumption, blood levels of glucose, lactate and pyruvate and their mutual relationship were discussed. From the practical point of view, it is important to arrange such a condition that newborns (including newborns who had suffered from intrauterine asphyxia and asphyxia) should not be allowed to cool down. (16 refs.) - *Journal summary*.

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2608 MORELLI, A. C., ACOSTA-FERREIRA, W., POLLINI, H., & DEL PINO, MARIA CLARA. Plasmatic protein separation by gel filtration chromatography and reproduction in vitro of fibres similar to the hyaline membrane of the newborn. *Biologia Neonatorum*, 14(3-4):142-152, 1969.

In a premature with an Rh problem, slight erythroblastosis and hyaline membrane disease, microscopic fibrin plus abnormal fibers (congo red dyed), irregularly shaped but in regular bundles and birefringent was found in some chromatographic fractions, after clotting with thrombin plus calcium. These appeared in fractions collected corresponding approximately to 170,000 to 1,050,000 mol.wt., mostly in the heavier weights. Future research will show whether they are normal to fetal life, prematurity, Rh problem, erythroblastosis or hyaline membrane disease. (19 refs.) - *Journal summary*.

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2609 JANCAR, J. Pellagra-like reaction due to PAS and isoniazid treatment. *Journal of Mental Subnormality*, 15(2):100-102, 1969.

Daily treatment of pulmonary tuberculosis in a 36-year-old, spastic, paraplegic MR with 15 gm of para-aminosalicylic acid (PAS) and 150 mg of isoniazid resulted in pellagra-like side effects which were reversed by withdrawing the PAS and isoniazid and administering

vitamin B, yeast, and nicotinamide daily. Initial treatment of tuberculosis also had included 1 gm of streptomycin daily for 12 weeks. The toxic effects of the PAS and isoniazid included cutaneous lesions, a gastro-intestinal disorder, peripheral neuropathy, and psychotic episodes. Temperature, blood pressure, and blood count remained normal. Blood urea was 31 mg/100 ml; serum cholesterol was 195 mg/100 ml; urine specimen showed normal concentration, 8% nitrogen, normal cystine, no keto acids reaction, no porphobilinogen, no protein precipitate, no reduction from the Benedicts test, normal calcium according to the Sulkowitch test, purple-black response (due to PAS) to the ferric chloride test, and a strong glycine pattern on the amino acid chromatogram. The pellegra-like symptoms abated after a few weeks of vitamin therapy (which was gradually reduced and discontinued after 2 1/2 years), and the tuberculosis lesion healed and remained static. (7 refs.) - A. Huffer.

Stoke Park Hospital Group
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2610 CRIST, TAKEY, & HULKA, J. F. Influence of maternal epinephrine on behavior of offspring. *American Journal of Obstetrics and Gynecology*, 106(5):687-691, 1970.

The daily subcutaneous injection of 0.4 mg epinephrine/kg into pregnant rats during the first 6 days of gestation decreased the weight of newborn rats; 6 of 10 pregnant rats treated with the catecholamine did not deliver. Further, the administration of epinephrine to pregnant rats on the seventh and twelfth days of gestation modified the behavior of their 23-day-old offspring; offspring

from treated mothers crossed less squares in open field tests and required more time to leave the home cages and reach food at the end of a passage after 24 hrs of food deprivation than controls. This suggested that epinephrine could produce fetal brain damage via placental vasoconstriction and secondary anoxia. (50 refs.) - K. Jarka.

School of Medicine
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Chapel Hill, North Carolina 27514

2611 BARROW, MARK V. Animal models in the clinical study of birth defects in man. In: National Foundation-March of Dimes. *The Clinical Delineation of Birth Defects. Part II. Malformation Syndromes*. (Birth Defects Original Article Series, Volume 5, Number 2.) New York, New York, 1969, p. 202-206.

Clinically produced malformation syndromes in animals may help in the delineation of teratogenically produced malformation syndromes in man. Phocomelia, micrognathia, and dislocations are seen in rhesus monkeys when thalidomide is administered from 24 to 30 days of gestation. Aortic lesions and enamel hypoplasia similar to that seen in hypercalcemic infants can be produced by an excess of vitamin D administered to rabbit embryos. Rats develop prognathism and limb dysplasia when subjected to excess vitamin A and a gibbous deformity of the spine when they receive β -amino-propionitrile *in utero*. Controlled prospective studies can be made and possible methods of prevention can be tested in these animal models. (13 refs.) - E. L. Rowan.

DEVELOPMENT ASPECTS - PHYSICAL

2612 HOFMEISTER, ALAN. Motor proficiency and other variables in educable mentally retarded children. *American Journal of Mental Deficiency*, 74(2):264-268, 1969.

This study was an investigation of the interrelations between motor proficiency and selected variables, including MA, school attainment, sociometric status, and classroom behavior in a selected sample of EMR boys and girls enrolled in a public school program. The results show motor proficiency to be related, in varying degree, to all the areas investigated, even when chronological and mental age factors were statistically partialled out. (12 refs.) - *Journal abstract*.

Department of Special Education
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2613 HAYDEN, FRANK J. The nature of physical performance in the trainable retarded. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 28, p. 219-226.

Correlations were computed between the physical performance score of TMR children (IQ range 20-55; CA range 8-18 yrs) and IQ and teacher ratings of emotional, mental, and social development. Physical performance variables were those usually associated with physical fitness as well as height, weight, muscle girth, external fat, balance, and swimming ability. Variables associated with organic or running-type endurance were more related to IQ than the other physical performance variables. Test-retest correlation coefficients (2- to 7-day retest period) for physical performance measures were consistently high, indicating that the performance of TMRs is relatively stable. Teacher ratings of psychological development were significantly related to physical performance scores, and teacher ratings of boys were significantly related to height, weight, and

age. There was a fairly high relationship between psychological development and 9 physical performance measures. Teacher ratings were most related to walking and running endurance. (No refs.) - J. K. Wyatt.

2614 KEOGH, JACK F., & OLIVER, JAMES N.

Physical performance of retarded children: Diagnosis and prescription. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 29, p. 227-231.

Since MR children are likely to exhibit less ability on physical tasks than other children, an individual diagnosis of their performance difficulties should be made, and appropriate remedial action prescribed. To prescribe for the MR child, the instructor must have detailed knowledge about specific movement problems or performance difficulties. The physical performance of 17 MR boys (CA 9-10) attempting to learn tasks involving the rhythmical use of limbs was observed. The problems and difficulties experienced by some boys were: interference between leg and arm movements; inability to perform a rhythmical pattern, control speed or force of movement, or perform in a limited area; the use of extraneous movements; the use of timid and indistinct movements; and task failure on one side of the body. Further analysis of the performance difficulties of the physically awkward child may lead to the identification of a syndrome of awkwardness which could be used as a basis for diagnosis and remediation (No refs.) - J. K. Wyatt.

2615 RARICK, G. LAWRENCE. The factor structure of motor abilities of educable mentally retarded children. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 31, p. 238-246.

An intercorrelation matrix of 16 physical growth and motor performance variables was used to determine the factor structure of the motor abilities of 9-, 12-, and 14-year old

EMR girls and boys. Factors common to girls and boys at these age levels were explosive muscular force, static strength, and coordination. An additional factor had consistent and high loadings of height and weight and seemed to be a factor of physique and body size. The factor structure of motor abilities of EMR children seems to be well-defined and relatively stable from 9 to 14 years. Between-sex differences were minor. These findings suggest that the organizational framework of motor abilities found in EMRs is similar to that of individuals with normal intelligence. (12 refs.) - J. K. Wyatt.

2616 STEIN, JULIAN U. Current status of research on physical activity for the retarded. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 32, p. 247-251.

Although research on the effects of physical education programs on the MR has been meager, available experimental and descriptive data indicate that positive physical, intellectual, and emotional benefits emerge from participation in regular, progressive, and systematic programs. Most studies have been descriptive and have involved institutionalized EMRs. Findings indicate that motor proficiency of MRs is below that of normals. However, the motor progress patterns of MRs and normals are similar, and MRs are nearer to physical than mental norms. MRs evidence higher achievement in activities which require simple neuromuscular skills and can learn motor skills as long as the practice provided is long enough for the acts to become neurologically grooved. EMRs who have participated in physical education programs have evidenced progress in academic subjects as well as increased self-concepts. (20 refs.)

2617 PILGRIM, DENNIS L., MILLER, FRANK D., & COBB, HENRY V. GSR strength and habituation in normal and nonorganic mentally retarded children. *American Journal of Mental Deficiency*, 74(1):27-31, 1969.

A group of 20 mildly MR children without known organic deficits, a second group of 20 normal children matched to the retardates on MA, and a third group of 20 normals matched to the retardates on CA were individually presented 20 short duration high intensity light stimuli. The independent variables were groups and trials, and the dependent variables were GSR (galvanic skin response) latency, amplitude, and duration. The results showed that all groups habituated to the

light presentations on all measures, and there were no significant differences in latency, amplitude, duration, or rate of habituation between the groups. (20 refs.)
Journal abstract.

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Vermillion, South Dakota 57069

2618 CLAUSEN, JOHS., & KARRER, RATHE. Temporal factors in autonomic responses for normal and mentally defective subjects. *American Journal of Mental Deficiency*, 74(1): 80-85, 1969.

Latency, time-to-peak, and recovery time were recorded for blood volume changes of the head, blood volume changes of the finger, and galvanic skin response (GSR) in response to a series of sensory stimuli. Three groups of 10 Ss each were included: normals, nonorganics, and organics. The only significant difference was for GSR recovery when this measure was adjusted for amplitude. The nonorganics had longer recoveries than the other groups. It was concluded that temporal factors do not discriminate between normal and mentally defective Ss as well as response frequencies do. (13 refs.) - *Journal abstract.*

Institute for Basic Research in
Mental Retardation
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2619 CAMPBELL, DONALD E. Circulorespiratory endurance of three age groups of institutionalized trainable mentally retarded males. *Training School Bulletin*, 66(2):60-65, 1969.

This investigation sought to evaluate circulorespiratory endurance of 3 age groups of institutionalized TMR males by means of endurance quotient. Three physical performance items, the 25-yard run for time, the 300-yard run for time, and the standing broad jump, were administered the 138 Ss. The time for the 300-yard run and the extended time for the 25-yard run were used as criterion measures to obtain the endurance quotient. This derived measure and the standing broad jump score were subjected to a single

classification analysis of variance for 3 subject groups. The conclusion is advanced that the circulorespiratory endurance of institutionalized TMR males does not vary by levels of chronological development. (12 refs.) - *Journal abstract.*

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2620 BAUMEISTER, ALFRED A., WILCOX, STEPHEN, & GREESON, JUDITH. Reaction times of retardates and normals as a function of relative stimulus frequency. *American Journal of Mental Deficiency*, 73(6):935-941, 1969.

Two studies compared young adult educable retardates and college students of comparable age on reaction time. In both studies, the Ss had to press a telegraph key in response to a red light and lift the key in response to a reaction signal (buzzer or light). A preparatory signal of 1.5 seconds preceded the reaction signal by 2, 4, 8, and 16 seconds (the 4 second interval was omitted in Experiment II). In both studies, normals were significantly faster and less variable. In Experiment I, presentation of the light or buzzer alone was compared to the presentation of light or buzzer in combination. Only on the light modality were responses slower when the light-buzzer combination was compared. In Experiment II, the probabilities of occurrence of both light and buzzer were altered (100% for each, 50% for each, and 10% and 90% for each). Slower reaction time was associated with decreased stimulus probability for both groups and for both types of signals. (11 refs.) - *J. M. Gardner.*

University of Alabama
University, Alabama 35486

2621 DeHAVEN, GEORGE E., MORDOCK, JOHN B., & LOYKOVICH, JOAN M. Evaluation of coordination deficits in children with minimal cerebral dysfunction. *Physical Therapy*, 49(2):153-157, 1969.

To develop a program of physical therapy for children with minimal brain dysfunction, movement coordination deficits associated with the disorder were delineated. A series of 14 tasks were administered to 122 children, 62 of whom were diagnosed as minimally brain damaged and the others as having normal cerebral function. Factor analysis revealed only the area of "primary deficits of distal alternate motion rate" as significantly

differentiating the 2 groups. The rates of finger wiggle, finger-thumb opposition, foot patting, and heel-toe walking with eyes closed reflect the dysfunction of small muscle groups in hands and feet characteristic of the "clumsy child". Therapy should be directed toward active and reciprocal exercise, progression of muscle participation from large to small muscle components, additional sensory stimulation, and emphasis on the coordination level of the individual child. (11 refs.) - *E. L. Rowan.*

Devereux Schools
Devon, Pennsylvania 19333

2622 BUTLER, BRUCE V., & ENGEL, RUDOLF. Mental and motor scores at 8 months in relation to neonatal photic responses. *Developmental Medicine and Child Neurology*, 11(1):77-82, 1969.

The relation of behavior (mental and motor) at 8 months to 3 physiological neonatal variables (birthweight, gestation age, and photic latency) was studied for 433 infants tested during the first 5 days of life. The group contained 285 whites (144 male, 141 female) and 148 blacks (72 male, 76 female). Photic latency refers to the interval between presentation of a light stimulus and the corresponding EEG responses in the occipital cortex. The 8-month behavioral measures were taken from a modified version of the Bayley scales in the areas of gross and fine motor skills and mental ability (responsiveness, vocalization, etc.). The correlations between all 3 neonatal measures and all 3 behavioral measures were small but significant. The highest correlation was between photic latency and mental ability ($r=.33$, $p<.001$), and the lowest was between birth weight and gross motor skills ($r=.12$, $p<.05$). When the other 2 physiological measures were partialled out, photic latency correlated $-.24$ with mental scores. (11 refs.) - *J. M. Gardner.*

University of Oregon Medical School
Portland, Oregon 97201

2623 AMES, LOUISE BATES. Children with perceptual problems may also lag developmentally. *Journal of Learning Disabilities*, 2(4):205-208, 1969.

Second graders from suburban public schools who were judged perceptually inadequate and immature on the Bender-Gestalt were also shown to lag developmentally on the Gesell Developmental Tests. A group (25) who received daily training in coordination and orientation for a period of 6 months improved

markedly in developmental tasks. Significantly, a control group of perceptually handicapped children without training fell even farther behind developmentally in the same 6-month period. Effective training can enable a child to perform at his highest potential developmental level, but it probably cannot speed his development. For the child who lags developmentally and does not repeat a grade or receive perceptual training, there is danger of further regression and inability to meet the demands of school and the outside world. (5 refs.) - E. L. Rowan.

Gesell Institute of Child Development
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- 2624 ZDANSKA-BRINCKEN, MARIA, & WOLANSKI, NAPOLEON. A graphic method for the evaluation of motor development in infants. *Developmental Medicine and Child Neurology*, 11(2):228-241, 1969.

Based on 34 stages of 4 different spheres of motor development (head and trunk, sitting position, standing position, and locomotion), grid of developmental age versus CA was constructed for each motor ability. The data were derived from monthly examinations of 212 Warsaw infants from the time they were 4 weeks of age to when they could walk by themselves. This method allowed simultaneous determination of different motor achievements. Deviation from the normal would indicate retarded or advanced motor development in either one or all spheres. (28 refs.)
L. S. Ho.

National Research Institute of
Mother and Child
Warsaw, Poland

- 2625 BEARGIE, ROBERT A., JAMES, VERNON L., JR., & GREENE, JOHN W., JR. Growth and development of small-for-date newborns. *Pediatric Clinics of North America*, 17(1): 159-167, 1970.

Follow-up study of 45 small-for-date infants at 13-53 months of age showed that 17 of them

remained undergrown (below the tenth percentile for their age) and 5 of the undergrown and 4 of the well-grown could not perform at their CA in 2 or more areas of development screening. The general health of all 45 children was good; however, most of them were slender and 6 were considered skinny. Congenital abnormalities were few. One child of the undergrown group had fixation nystagmus and another deafness due to rubella. Abnormalities of the well-grown group included severe genu valgum, micrognathia, and esotropia. One third of the families were of marginal socioeconomic status. The eventual height of a child is primarily determined genetically; however, maternal complications occurring before the third trimester are likely to induce permanent stunting in growth (8 refs.) - L. S. Ho.

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Lexington, Kentucky 40506

- 2626 EDWARDS, NANCY. The Relationship Between Physical Condition Immediately After Birth and Mental and Motor Performance at Age Four. In: *Genetic Psychology Monographs, Volume 78*. Provincetown, Massachusetts, Journal Press, 1968, p. 257-289.

The physical condition immediately after birth as evaluated by Apgar ratings at 1-minute and 5-minutes of age and observations at 20 minutes of age of 147 infants was correlated with Stanford-Binet Intelligence Scale, concept formation test (Graham-Ernhart Block Sort), fine motor tasks (Porteus Maze, Wallin Pegboard, copying forms, and stringing beads), and gross motor tasks (line walk, hopping, and ball catching) at age 4. There was a highly significant relationship ($p < .001$) between neonatal Apgar scores and the 4-year measures. The 5-minute Apgar score accounted for more of the variance of 4-year development than the other neonatal measures; it contributed more than twice as much weight as the 1-minute score. Physical condition at 1 and 5 minutes correlated higher with fine and gross motor coordination of age 4 than with IQ and concept formation at age 4 ($r = .400$ to $.480$ compared with $.251$ to $.320$). Significant relationships between the 20-minute examination and the 4-year measures and between the neonatal measures and the quality of performance and emotional rating at 4 years of age were also found; birth weight was not related to the 4-year measures. (26 refs.) - M. G. Conant.

DEVELOPMENTAL ASPECTS - MENTAL

- 2627 LOGAN, DON R. Paired associate learning performance as a function of meaningfulness and response times. *American Journal of Mental Deficiency*, 74(2):249-253, 1969.

The paired-associate learning and retention performance of 80 MR Ss was investigated using 2 response time conditions and 2 levels of meaningfulness. Analysis of original learning, immediate recall, and 24-hour relearning data revealed that both main effects were significant. Analysis of 24-hour recall demonstrated significance for the main effect of meaningfulness but not for response time. The interaction effect was not significant on any of the 4 measures. (17 refs.) - *Journal abstract*.

University of Utah
Salt Lake City, Utah 84112

- 2628 KELLAS, GEORGE, & BAUMEISTER, ALFRED A. Response learning and the paired-associate performance of mental retardates. *American Journal of Mental Deficiency*, 74(2):273-278, 1969.

Two investigations were conducted to determine whether a model of paired-associate (PA) learning derived from normal adults applies to the PA acquisition of retardates. In the first study, either relevant or irrelevant pretraining was provided prior to a PA task. Analyses of the PA data indicated that prior familiarization with relevant responses benefited PA learning. The second experiment examined the progress of response learning within PA acquisition. The results showed the free-recall of the responses was initially superior to PA recall. These results were in agreement with a model advanced by Underwood and Schulz (1960). (6 refs.) *Journal abstract*.

University of Kansas Medical Center
3933 Eaton Street
Kansas City, Kansas 66103

- 2629 HAWKER, JAMES R., & KEILMAN, PEGGY A. Prompting and confirmation in paired-associate learning by retardates. *American Journal of Mental Deficiency*, 74(1):75-79, 1969.

Forty retarded Ss learned an 8-item paired-associate list to a criterion of 2 consecutive errorless test trials, and returned one week later to relearn the list to criterion. A 2x2 factorial design was employed in which the 2 variables were training procedure (Prompting or Confirmation) and response availability (8 or 16 total response terms). Training procedure did not differentially affect acquisition during original learning, but the relearning and retention data both indicated significantly better retention following a Confirmation procedure. Increasing the total number of response terms used in making up alternatives facilitated acquisition during the original learning session, but did not influence retention or relearning. (10 refs.) - *Journal abstract*.

Lamar State College of Technology
Beaumont, Texas 77705

- 2630 SIEGEL, PAUL S., & SCHNEIDER, ROBERT. Discrimination learning of retardates in relation to incentive motivation and attention. *Journal of Comparative and Physiological Psychology*, 68(4):656-658, 1969.

The role of attention and incentive motivation in relation to learning a discrimination task was studied in 75 institutionalized MRs (CAs 9-36 yrs; IQs 45-60). Etiology was not controlled. Of the 46 males and 29 females who were pretested for position bias, 5 were discarded; also 2 were discontinued because they showed an aversion to the rewards. Four groups were employed and matched in terms of CA, IQ, sex, and previous lab experience. One group was reinforced for success on a 2-choice simultaneous junk-object discrimination with a penny on one side and a washer on the other; the second group was reinforced with candy corn on one side and chiclet gum on the other; the third group received chiclet gum on both sides; and the fourth group received candy corn on both sides. For the

purpose of analysis, the last 2 groups were combined into one identical reward group. The Modified Wisconsin General Test Apparatus was used, and each S was given 32 trials a day for 5 days or run to a criterion of 12 successive correct responses. The data indicated that differences in incentives affected learning. Group 1 had the poorest performance, while groups 3 and 4 scored the highest. Attention and incentive are important variables in learning this discrimination task. (5 refs.) - B. Bradley.

University of Alabama
University, Alabama 35486

2631 BRICKER, WILLIAM A., HEAL, LAIRD W., BRICKER, DIANE D., HAYES, WILLIAM A., & LARSEN, LAWRENCE A. Discrimination learning and learning set with institutionalized retarded children. *American Journal of Mental Deficiency*, 74(2):242-248, 1969.

A 2x2 analysis of variance design was used and included the dimensions of fading versus no fading and position-extinction versus no extinction. Sixteen institutionalized retarded children were assigned to 1 of the 4 treatment groups and were given 96 training problems and 32 test problems. The results indicated each of the 3 experimental procedures produced fewer errors across problem blocks than did the control procedure. Analysis of the test data indicated the experimental procedures facilitated the learning of individual problems but not the acquisition of learning set. (13 refs.) - *Journal abstract*.

Peabody College
Nashville, Tennessee 37200

2632 SEITZ, SUE. The effects of variations in confirmation training on discrimination performance. *Psychonomic Science*, 14(4):145, 147, 1969.

Performance of 24 institutionalized EMRs on the confirmation training procedure improved significantly when interspersed test trials were eliminated. The Ss were randomly assigned to 3 groups which did not differ significantly in IQ or CA. An automated teaching device presented 8 pairs of pictures randomized for presentation and position order. All Ss were required to learn that the correct response was one member of each pair. For Group 1, 20 test trials were interspersed with 20 practice trials with the S's responses advancing the program. Ss in Group 2 were reinforced (candy and verbal)

by the experimenter on only correct practice trials; the program advanced automatically after each response on practice and test trials. For Group 3, the experimenter reinforced (candy and verbal) each correct response and advanced the program by remote control; the 20 practice trials also served as test trials, and a response was recorded only when the first choice was correct. Error scores for the first and last blocks of trials were compared. Analysis revealed no significant difference between Groups 1 and 2; however, when these groups were compared with Group 3, the latter showed significantly fewer errors ($p < .01$). The comparability of prompting and confirmation as alternative training procedures should be reconsidered. (7 refs.) - A. Buffer.

Austin State School
Austin, Texas 78767

2633 MASSEY, PHILLIP S., & INSALACO, CARL. Aversive stimulation as applied to discrimination learning in mentally retarded children. *American Journal of Mental Deficiency*, 74(2):269-272, 1969.

This study investigated the relationship between the discriminative aspects of aversive stimulation and discrimination learning in institutionalized retarded children. Four matched groups were given various combinations of candy reinforcers and aversive stimulation in a simple discrimination learning task. The results demonstrated that aversive white noise can, under certain conditions, aid acquisition of a response. Its greatest facilitating effect is when it functions as a cue for an incorrect response. In addition, when associated with a correct response, aversive stimulation also facilitates acquisition. (17 refs.) - *Journal abstract*.

Habilitation Center
Ladson, South Carolina 29456

2634 VAN OUDENHOVEN, N. J. A., & VAN DER AART, W. J. P. Auditory discrimination learning and transfer in imbeciles. *Journal of Mental Subnormality*, 14(27, Part 2): 98-100, 1968.

A study involving 20 TMRs (CA 16-27 yrs; MA 39-55 mos) provides evidence that results obtained in visual discrimination and transfer studies can be generalized to auditory discrimination fields. Two groups of 10 Ss were given pictures of 5 instruments to observe while they listened to the music of the

different instruments. The Ss were instructed to push the button beneath the appropriate picture when they heard that instrument. Findings support the conclusions that poor discrimination learning in MRs is caused by a failure to attend to relevant stimulus dimension, discrimination learning remains on a chance performance level for several trials, then improvement occurs relatively fast, and transfer is rather impressive and caused by the creation of "learning set" and increased conceptual and perceptual abilities. Important educational considerations include reducing stimuli and providing an opportunity to acquire clear S-R connections. (4 refs.)
G. M. Nunn.

Leiden University
Leiden, Netherlands

- 2635 AUXTER, DAVID. Effects of reinforcement on motor learning and retention by mentally retarded. *Perceptual and Motor Skills*, 29(1):99-104, 1969.

The effects of reinforcement on learning and retention and patterns of motor learning were studied. Each of 2 groups of MR children (N=16/group), matched for IQ and CA, were initially given 25 trials on a stabilometer. After 6 months, 5 retention trials were given. One group was reinforced with candy on trials 5 to 25. The other group was not reinforced. The results show that MR children are capable of learning motor tasks involving rapid motor adjustments; reinforcement of a gross motor task may facilitate greater learning and it retards the effects of the onset of satiation, which leads to decrements in performance and reinforcement for performance on the stabilometer does not result in significantly greater retention over a 6-month interval. (11 refs.) - *Journal abstract*.

Slippery Rock Pennsylvania State College
Slippery Rock, Pennsylvania 16057

- 2636 GALLAGHER, JOSEPH W. Short-term recall of sentences in normal and retarded children. *American Journal of Mental Deficiency*, 74(1):57-61, 1969.

The present study employed 2 groups of normal children and 2 groups of retarded children. Each S was presented with a series of sentences. The S was asked to recall 3 linguistically different sentences after either a 0, 8, or 15 second delay. The results indicated that the older normal group and the matched MA retardate group performed similarly

on all 3 types of sentences at 0 second delay. With the 2 longer delay intervals, the older normals performed better than the matched-MA retardates on more complicated sentences. Comparisons between the young normal and matched-MA retardate groups indicated that the young normals performed better than the matched retardates at all intervals and for all sentence types. (23 refs.)

Journal abstract.

University of Alabama
University, Alabama 35486

- 2637 RYAN, J. F., CHIVERS, J., & REDDING, G. Short-term memory and rehearsal in educable subnormals. *American Journal of Mental Deficiency*, 74(2):218-222, 1969.

Short-term memory in educable subnormal adolescents was investigated by comparing their recall of ungrouped and of temporally grouped sequences. It was argued that if the stimulus trace theory of mental retardation (Ellis, 1963) is correct, then no improvement in recall with grouping would be expected. However, the subnormals did improve their recall and also showed all the other effects characteristic of normals. The implications of this are discussed. An alternative explanation of short-term memory defects is proposed in terms of a failure of spontaneous verbal rehearsal. (12 refs.)

Journal abstract.

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5, Salisbury, Villas
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- 2638 ELLIS, NORMAN R. Short-term memory and rote learning in retarded and normal subjects. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 20, p. 150-163.

Marked differences in short-term memory (STM) processes were found in a group of 21 seemingly homogeneous, mildly retarded Ss (CAs 16-24 yrs; IQs 57-70; 6 females, 15 males). STM tests used were delayed matching-to-sample, a children's card game, and digit span. Rote learning tests with meaningful and nonmeaningful cues were also administered. The performances of the MR Ss (MA 9-10 yrs) on the children's card game test were at a lower level than those of normal Ss (CA 4-5

yrs) in a previous study. The only STM measure which was related to rote learning was the children's card game, and this was only on the meaningful task. There were moderate correlations (range .42 to .62) between the delayed matching to sample test, the children's card game, and the digit span test (3 refs.) - J. K. Wyatt.

2639 RASKIN, LARRY M. Long-term perceptual memory in normal and educable retarded children. *American Journal of Mental Deficiency*, 73(6):903-905, 1969.

The effect of prior training on the perception of apparent movement was studied in younger (CAs 8-3 to 13-0) and older (CAs 11-0 to 15-2) retardates and equal-MA school children (first and fourth graders). The experimental task involved presentation of 30 consecutive trials of two 3 1/2 inch arrows separated by a distance of 3 1/2 inches. The arrows were presented successively to give the illusion of apparent movement. Prior to this task (24 hours), each S performed a similar task with an arrow and a square as the stimuli. In the first test (arrow and square), neither the retardates nor the normals perceived movement. In the second test (arrow and arrow), the younger retardates reported seeing movement more often than their equal-MA normals ($\chi^2=4.89$, $p<.05$). In the older Ss, 9 of the 15 retardates and 3 of the 15 normals reported movement; however, this difference was not significant. It is concluded that prior training interferes with the perception of apparent movement for normals but not retardates, who may require more training sessions to equal the same memory efficiency as normals. (16 refs.) - J. M. Gardner.

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2640 ARGRANOFF, B. W. Memory formation in the goldfish. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 16, p. 131-133.

Studies of learning in the goldfish indicate that memory formation requires specific molecular processes and that these processes can be chemically dissociated from other behavior. Experiments in which intracranial injections of antimetabolites were given either prior to or at various times after

training on a shock avoidance task suggest that goldfish have short-term and long-term memory which is not affected by puromycin. However, puromycin appears to selectively block the formation of long-term memory. Chemical studies show that protein synthesis is inhibited following the injection of either puromycin or acetoxycycloheximide. Since the behavioral effects of puromycin last for a shorter period of time than the inhibition which occurs when leucine ³H is incorporated into brain protein, it may be that the chemical process involved in memory fixation involves a protein which recovers more rapidly from the puromycin block than most proteins. Experiments on the manipulation of the period of puromycin susceptibility in newly-trained goldfish suggest that memory fixation begins when arousal state decreases. (7 refs.) - J. K. Wyatt.

2641 FOLLINI, PAUL, SITKOWSKI, CAROLYN A., & STAYTON, SAMUEL E. The attention of retardates and normals in distraction and non-distraction conditions. *American Journal of Mental Deficiency*, 74(2):200-205, 1969.

Thirty normals and 29 retardates, approximately equal in CA, were compared in their ability to concentrate on metronome beats of varying rates in 2 situations, one where a distracting beat was present and one in which there was no distracting beat. In the non-distraction condition there was little difference between normals and retardates in their estimate of focal beat rates. While there was somewhat more difference between the groups in the distraction condition, the main finding was that retardates' concentration on the focal beat was significantly more functionally related to the distracting tempo than was that of the normals. (5 refs.) - *Journal abstract*.

Belchertown State School
Belchertown, Massachusetts 01007

2642 MacMILLAN, DONALD L. Motivational differences: Cultural-familial retardates vs. normal subjects on expectancy for failure. *American Journal of Mental Deficiency*, 74(2): 254-258, 1969.

An interrupted task paradigm was used to determine whether cultural-familial retardates exhibited a higher expectancy for failure than non-retarded Ss. A total sample of 120 Ss included equal numbers of: primary retardates, intermediate retardates, and non-retarded Ss. Data analyses revealed that

both retarded groups placed blame on themselves for the tasks not being completed. Resumption choice and placement of blame were significantly related for non-retarded Ss but not for retardates. Results were interpreted to support the notion that cultural-familial retardates do have a higher expectancy for failure than do normals. (11 refs.)

Journal abstract.

University of California
Riverside, California 92502

2643 SEITZ, SUE, GOULDING, PEGGY, & CONRAD, ROBERT W. The effects of maturation on word associations of the mentally retarded. *Multivariate Behavioral Research*, 4(1):79-88, 1969.

A 30-month follow-up study of the performance of 53 institutionalized adolescent MRs on a word association test was designed to test Lenneberg's hypothesis that change in language functioning does not occur after the fourteenth year and Moran's hypothesis that associative sets proceed in developmental sequence from concrete to abstract. The group who originally had high MAs (N=20; mean MA=11.5) did not change significantly in terms of type of responses or reaction time. The low-MA group (NA=33; mean MA=7.6) showed significant increases in abstract associations ($t=6.99$, $p<.001$) and reduction in reaction time ($t=6.99$, $p<.001$)--both hypotheses were supported. (12 refs.) - J. M. Gardner.

Austin State School
Austin, Texas 78767

2644 KINTZ, B. L., FOSTER, MARILYN S., HART, JAMES O., O'MALLEY, JOHN J., PALMER, EDWARD L., & SULLIVAN, SHARON L. A comparison of learning sets in humans, primates, and subprimates. *Journal of General Psychology*, 80(2):189-204, 1969.

Review of research on discrimination learning sets suggests that they exist along an interspecies continuum and that the differences between the species are quantitative as well as qualitative. Results in human studies show that discrimination learning sets are present in normal and MR children. Level of intellectual functioning correlates positively with speed and efficiency of learning set in normal and MR children when the 2 groups are compared. Data from animal studies show that learning set formation in primates seems to follow phylogenetic lines with quantitative and qualitative variations. Research studies relating to subprimates show that

they also form learning sets with efficiency decreasing according to level of phylogenetic scale. Learning sets in rats require more extensive research, but preliminary data suggest that the rat may be able to develop learning sets. One finding indicates that interproblem learning curves for normals and retardates "accelerated quickly from the outset making it difficult to discern an S-shaped pattern." (50 refs.) - B. Bradley.

Western Washington State College
Bellingham, Washington 98225

2645 BUTTERFIELD, EARL C., & MCINTYRE, ANNE. Cognitive and motivational factors in concept switching among the retarded. *American Journal of Mental Deficiency*, 74(2):235-241, 1969.

Two studies evaluated the effects of the intellectual variables MA, and IQ, and the experiential variables, social deprivation (SD) and length of institutionalization (LI), upon the concept switching of MR Ss. The first study indicated that MA, IQ, and SD significantly and independently discriminated between groups constituted on the basis of their concept switching behavior. The second study showed that 2 different types of punishment facilitated the concept switching behavior of retardates. It was concluded that both cognitive and motivational variables contribute to concept switching. (20 refs.) - *Journal abstract.*

University of Kansas Medical Center
Kansas City, Kansas 66103

2646 KAUFMAN, MELVIN E., & GARDNER, WILLIAM I. Transfer of training of learning sets in mental defectives: I. Discrimination reversal. *American Journal of Mental Deficiency*, 73(5):801-803, 1969.

The nature and direction of transfer of training of an object-quality learning set (LS) to a discrimination-reversal LS were studied with 26 noninstitutionalized mildly retarded school children. Twelve experimental Ss received object-quality discrimination prior to reversal learning, and 14 control Ss received reversal problems only. A modified Wisconsin General Test Apparatus was used, and the stimuli were drawn from a pool of 250 pairs of objects differing in size, shape, and color. In Phase I of the object-quality LS training (150 trials), each problem was run to a criterion of 9 of 10 correct responses. In Phase II, all Ss received 50 3-trial

problems, and only those Ss meeting a 90% correct trial-2 response were included. In the reversal test, 8 problems were presented daily for 5 consecutive days. Verbal instructions were kept to a minimum, and each S was adapted to the apparatus for 10 minutes. The experimental Ss did significantly better on the second and third post-reversal trials (the first post-reversal trial served to let the Ss know that a shift in reward was made). On the second post-reversal trial, 7 of 12 experimental Ss as compared to only 1 of 14 control Ss reached the criterion of 90% correct. On the third post-reversal trial, none of the control and one-half of the experimental Ss reached criterion. This clearly indicated positive transfer of training for the experimental Ss. (6 refs.) - J. M. Gardner.

University of Wisconsin
Madison, Wisconsin 53703

- 2647 SMITH, JEROME, & TUNICK, JEFFREY. Cross-modal transfer of a discrimination by retarded children. *Journal of Experimental Child Psychology*, 7(2):274-281, 1969.

Transfer of a discrimination from visual- to tactual-kinesthetic, and from tactual-kinesthetic to visual sense modes was examined in a miniature experimental design which employed a modified Wisconsin General Test Apparatus. Ss were 6 institutionalized male MRs (mean CA 147 mos; mean MA 85 mos; mean IQ 58). The 24 stimulus objects were 6 plastic geometric forms of equal volume and 4 textures. Correct responses were reinforced with candy and a verbal "good", and incorrect responses were followed by a verbal "no". Ss were selected for testing on the basis of their performance on pretraining problems. In the experiment, each S served as his own control and was given 12, 4-trial problems daily for a 30-day-period. Variables of type of transfer and dimension relevant to solution were manipulated. When cues for both modalities were identical, Ss evidenced significant cross-modal transfer ($p < .001$); however, transfer did not occur when only information about the relevant dimension was available. Ss exhibited a significant ($p < .025$) preference for texture as opposed to form relevant. These data indicate that MR children can transfer across sense modes; however, they do not support a hypothesis of dimension-transfer. (12 refs.) - J. K. Wyatt.

University of Connecticut
Storrs, Connecticut 06268

- 2648 SHIELDS, RUTH V., *GORDON, MARY ALICE, & EVANS, SELBY H. Schematic concept formation in relationship to mental ability in adolescents. *Psychonomic Science*, 17(6):361-362, 1969.

The California Test of Mental Maturity (CTMM) IQ scores of 60 Ss (mean CA 16 yrs) divided into 3 IQ groups (MR, 70-85; average, 90-109; superior, 120-140) correlated positively ($r = .36$) with schematic concept formation (SCF) scores. The Ss were asked to make dichotomous (similar-different) judgments on 60 pairs of stimuli (patterns of numbers) which had been generated by the VARGUS 9 computer system and which were examples of 2-schema families at 70% redundancy. Correlation between the language section of the CTMM and the SCF was .28, while correlation between the nonlanguage section of the CTMM and the SCF was .42. The difference between these correlations was significant ($p < .05$). No significant differences between the male and female test scores were found. The SCF has potential utility in studies of individual differences because the task is largely non-verbal, the stimuli are relatively unfamiliar thus a S's performance depends on what he can learn on the task itself, and the levels of difficulty and other characteristics of the task can be quantified. (17 refs.) - A. Huffer.

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- 2649 RYBOLT, GAYLORD A. Stability characteristics of the semantic differential responses of the mentally retarded. *Psychological Reports*, 24(1):103-108, 1969.

An investigation of the stability or consistency of the responses of MRs to 2 semantic differentials--the Semantic Rating Scale (SRS) and the Reliability Scale (RS)--revealed a significant relation between response consistency and intelligence ($p < .001$) and no significant deviations on immediate, 1-week, or 3-week retest ratings. Ss were 79 EMR students (verbal IQ range 50-85; CA 13 to 17 yrs) who attended public school special education classes. The SRS was composed of 5 concepts which were rated on 25 bipolar scales, and the RS consisted of the same 5 concepts and 10 scales which had been randomly selected from the SRS. Each scale had 5 response categories. Retest data were obtained for 73 Ss. There were no significant differences in stability as a function of time between testing. Reliability coefficients ranged from .31 to .82 for the 10

scales. The responses of relatively more intelligent Ss were more consistent than those of relatively less intelligent Ss. (7 refs.) - J. K. Wyatt.

University of Oregon
Eugene, Oregon 97201

2650 REIHER, ROBERT H., PHELAN, JOSEPH G., & KIKER, VERNON L., JR. Reversal and non-reversal shifts in neurologically handicapped children. *Journal of Psychology*, 72(1):41-53, 1969.

The NYU Card Sorting Test was used to investigate the relative effectiveness of a reversal shift as compared with a nonreversal shift in 24 neurologically handicapped children (CA range 6 to 15 yrs). The concepts were shape and color, and the response measure was number of trials. A 2 X 2 factorial design was employed in which 2 nonreversal groups learned either Shape-Reverse Color or Color-Reverse Shape and 2 reversal groups learned either Shape-Reverse Shape or Color-Reverse Color. A control group, whose training began with the first stage of the second concept, was used to investigate transfer effects. There were no significant differences between total shape and total color groups; between-group differences on the learning of the second concept were slight and insignificant. These findings were similar to those for brain-injured adults. Although there were no significant differences between reversal and nonreversal groups on the first stage of learning of the second concept, their range of scores was much higher than that of normal adults and similar to that of brain-injured adults. The performance of neurologically handicapped control group Ss evidenced a lack of transfer effects. The reversal shift did not occur at a significantly faster rate than the nonreversal shift; data again are consistent with those for brain-injured adults and different from those for normal adults. These data suggest that neurologically handicapped children may have an impairment in the mediational process which facilitates reversal shift learning in the normal adult. (11 refs.) - J. K. Wyatt.

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2651 SCHWARZ, ROBERT H., & JENS, KENNETH G. The expectation of success as it modifies the achievement of mentally retarded adolescents. *American Journal of Mental Deficiency*, 73(6):946-949, 1969.

The effects of special class placement and test instructions on an intelligence test task (similar to the coding subtest of the Wechsler scales) were examined in 40 EMR adolescents. The Ss were divided into 4 groups: 2 groups were tested in the special class setting and 2 groups were integrated with regular students. The groups were further divided according to whether test-taking instructions emphasized intelligence (I) or coordination (C). It was predicted that the Ss would be sensitive to past failures and that their performance would be influenced by the presence of a peer group known to be brighter. Thus, highest scores were predicted for the special class group tested under the C condition, and lowest scores for the regular class tested under the I situation. A 2 X 2 analysis of variance revealed that the special class group did significantly better than the peer group ($F=8.69$, $p<.01$); however, there were no significant differences between instructions. In fact, under the peer group conditions, Ss did better under the I situation than under the C situation which was contrary to predictions. This finding suggests that the Ss may have been more motivated to avoid failure than to achieve success. (9 refs.) - J. M. Gardner.

University of Wisconsin
Madison, Wisconsin 53706

2652 KOPATIC, NICHOLAS J., & KOPATIC, NEDA. The reliability of mental retardates in judging subjective phenomena--Part II. *Training School Bulletin*, 66(2):86-88, 1969.

Auditory differential thresholds were obtained on a normal and retarded sample of Ss by using the method of constant stimuli. The results of the study confirm previous findings which showed that MR Ss are not as reliable as normal Ss in reporting auditory differential thresholds. The significance of this finding is discussed. (3 refs.) - *Journal Abstract*.

Cumberland County School
Bridgeton, New Jersey 08302

- 2653 ALLEN, ROBERT M., & WALLACH, EDWARD S. Word recognition and definition by educable retardates. *American Journal of Mental Deficiency*, 73(6):883-885, 1969.

The difference between the ability to identify objects by name (word recognition) and the ability to define those names (word definition) was examined with 50 moderately and mildly retarded institutionalized children. The Peabody Picture Vocabulary Test Form B was used. After each S had been asked to define words which represented pictures from the test, they were given the actual test. In both situations, testing continued to a criterion of 6 of 8 consecutive failures. No attempt was made to vary the sequence of administration. Word recognition was significantly greater ($p < .001$) than word definition when the ceiling (highest correct response) and the total number of correct responses were examined. Only 7 of the 50 Ss correctly defined words which they had failed to recognize. This study indicates that verbal encoding is a major weakness in EMRs. (9 refs.) - J. M. Gardner.

University of Miami
Coral Gables, Florida 33124

- 2654 SONG, A. Y., & SONG, R. H. Visual memory and reading ability of mental retardates. *American Journal of Mental Deficiency*, 73(6):942-945, 1969.

The relative contribution of visual-motor ability (VM), VM ability under memory conditions, and visual memory to reading ability was examined in high ($N = 26$) and low ($N = 28$) reading institutionalized retarded adolescents equated for MA, CA, and IQ. The groups were divided according to scores above or below the 2-year median on the Wide Range Achievement Test. VM ability was tested by using the Bender-Gestalt (scored with a modified Pascall-Suttell system). VM memory was scored by having the Ss study the Bender cards for only 10 seconds and then reproduce them (under normal conditions the card is left available). Visual memory was measured by subtracting scores achieved on the first test from those on the second test. The only significant difference between the 2 groups of readers was on the visual memory index: the high reading Ss did significantly better ($t = 2.13$, $p < .025$, one-tailed). Both groups scored lower under the VM memory condition

than under the standard condition. This study emphasizes the importance of immediate visual memory in reading skills for the retarded. (16 refs.) - J. M. Gardner.

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- 2655 BEIER, ERNST G., STARKWEATHER, JOHN A., & LAMBERT, MICHAEL J. Vocabulary usage of mentally retarded children. *American Journal of Mental Deficiency*, 73(6):927-934, 1969.

The vocabulary word usage of 30 institutionalized male MRs (CAs 11-24 yrs; IQs 23-75) was compared to normative data available on normal 12- and 16-year olds. The MRs were selected on the basis of their ability to communicate. Word usage samples of 2,700 words were tape recorded over a 2-hour period. Various stimulus words (girls, sex, war) were used to stimulate conversation. The retardates were found to speak significantly more slowly ($t = 11.7$, $p < .01$), refer less often to others ($t = 5.4$, $p < .01$), and use more self-references ($t = 7.0$, $p < .01$) than normals. Brighter retardates spoke a little faster, used more self-references, and asked more questions. Lower level Ss used more different words; however, this was a function of poor sentences in which objects tended to be enumerated. Usage of the 10 most frequently used words (and, I, you, the, it, to, a, is, not, that) was similar for normals and retardates. (16 refs.) - J. M. Gardner.

University of Utah
Salt Lake City, Utah 84112

- 2656 MARTYN, MARGARET M., *SHEEHAN, JOSEPH, & SLUTZ, KAREN. Incidence of stuttering and other speech disorders among the retarded. *American Journal of Mental Deficiency*, 74(2):206-211, 1969.

A cross-validating sample of 346 MR patients at Camarillo State Hospital, California, revealed the incidence of stuttering to be the same as that for the population generally. A recent survey at Porterville State Hospital, California, by Sheehan, Martyn, and Kilburn (1968) reported a similar incidence figure. Patients from 8 wards at Camarillo were given individual diagnostic speech evaluations. Twenty percent of those examined had normal speech. One-third had no speech or delayed

speech, while more than one-third had articulatory disorders. Among the remainder were 12 voice problems, 3 stutterers, 1 aphasic, 1 hearing loss, 1 cerebral palsy, and 1 clutterer. (21 refs.) - *Journal abstract.*

*University of California
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- 2657 KAPPELMAN, MURRAY M., KAPLAN, EUGENE, & GANTER, ROBERT L. A study of learning disorders among disadvantaged children. *Journal of Learning Disabilities*, 2(5):261-268, 1969.

While the effects of cultural and educational deprivation are obvious determinants of learning disorders among disadvantaged children, the effects of organic involvements are often overlooked. A multidisciplinary team evaluated 506 children referred to a health center because of learning problems. Evaluation procedures involved: school reports; parent and child interviews; pediatric and neurologic examination; and psychological, language, and articulation testing. Of the 306 children diagnosed as having significant learning disabilities, 55% had organic handicaps. Cultural deprivation was considered the major agent for only 6% of the children, while emotional problems were the principal cause in approximately 1/4 of the Ss. It is important to recognize that the conditions of poverty often result in high rates of maternal and perinatal complications which can lead to organic deficiencies. (6 refs.) - *J. M. Gardner.*

Sinai-Druid Comprehensive Health Center
Baltimore, Maryland

- 2658 RUBIN, SUELLEN SAFIR. A reevaluation of figure-ground pathology in brain damaged children. *American Journal of Mental Deficiency*, 74(1):111-115, 1969.

Brain damaged children were compared with familial retardates and normal children on their perception of tachistoscopically presented stimuli. When tested under minimally distracting conditions, the brain damaged Ss showed no significant difference from the controls in their ability to recognize figure-ground configurations. When forced to choose between either the correct figure or the correct ground, brain damaged Ss and controls alike usually chose the correct figure. This experiment raises questions about Werner and Strauss's earlier finding of

figure-ground pathology in brain damaged children and discusses their findings in terms of possible alternative causes. (7 refs.) - *Journal abstract.*

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- 2659 WINTERS, JOHN J., JR., & GERJUOY, IRMA R. Recognition of tachistoscopically exposed letters by normals and retardates. *Perception and Psychophysics*, 5(1):21-24, 1969.

The effects of varying the length of tachistoscopic exposure were examined in 30 institutionalized EMRs (mean CA = 15.8; mean IQ = 64.2; mean MA = 10.06) and normals with equal CA and MA. The stimulus consisted of cards (25 cm x 27 cm) containing 4 vowels (A,E,O,U) randomly placed. Dark adaptation and pretest training were done for each S. Exposure times were 0.1 and 0.3 seconds. Inter-trial intervals were 20 seconds. The Ss were asked to recognize as many letters as possible. No significant differences were noted between males and females. Under the longer exposure time, all 3 groups performed better. Under the shorter time, there were no differences between the groups; however, under the longer time, the equal CA group performed best, and the retardates recognized the fewest letters. Right-field recognition was superior under the short exposure for the MRs and equal CA normals, while there were no significant differences under the longer exposure. (10 refs.) - *J. M. Gardner.*

Edward R. Johnstone Center
Bordentown, New Jersey 08505

- 2660 JONES, M. H., DAYTON, G. O., LIMPAECHER, R., MURPHY, T. V., & HIRSCH, P. Electrooculographic studies in cerebral palsied and in normal children. In: Gardiner, Peter, MacKeith, Ronald & Smith, Vernon, eds. *Aspects of Developmental & Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p. 48-58.

The ability of 20 Ss (6 normal children, 3 normal adults, 10 infants with cerebral palsy (CP), 1 CP boy, and a 30-month-old child with congenital nystagmus) to fix and follow a moving target was determined by electro-oculography, tape recordings, and computer programming. The targets consisted of 2-inch Fantz faces and 1 1/2-inch diameter simple

colored pictures which were placed on translucent paper that was moved at a speed of 16°/sec. Calculations indicated that left to right following is better than right to left in the central 20° of the follow movement (LR 98-99% in adults); infants 25 and 26 months old showed no consistency. The data obtained from normal children are close to that of adults in linear correlation of each eye with target and in conjugation of both eyes in pursuance of a moving object. Young CP children with normal motility in routine ophthalmological tests had poorer conjugation and greater deviation of each eye from target (as measured by electro-oculography) than normal children. (8 refs.) - V. G. Votano.

in the case of Turner's syndrome where IQ may vary from < 70 to > 130 and perception and conceptualization of space-form relations may be faulty. In hypoparathyroidism and pseudohypoparathyroidism, the intelligence level may be depressed; however, in other endocrine defects including hyperthyroidism, hypopituitary dwarfism, Addison's syndrome, and Cushing's syndrome, the intelligence is not appreciably affected. (34 refs.) - L. S. Ho.

- 2661 GARDNER, THOMAS D., & BARNARD, JAMES W. Intelligence and the factorial structure of person perception. *American Journal of Mental Deficiency*, 74(2):212-217, 1969.

This study investigated the relationship between intelligence and the factorial structure of person perception. Ten retardates of secondary school age, 10 sixth-graders of average intelligence, and 10 gifted fourth-graders were the Ss. Each was asked to rate a series of 34 pictures from the Frois-Wittman series on 15 adjectival scales, each being bipolar and representing a different affect. The groups were significantly different ($p < .05$) on 4 analyses of the principal-axis factor analysis and Procrustes rotation: total variance accounted for by the factor analysis, size of the first factor, hyperplane count, and congruence between the obtained principal-axis solution and a target matrix. It was concluded that intelligence is a determinant of person perception. (9 refs.) - *Journal abstract*.

University of South Florida
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- 2662 MONEY, JOHN. Intellectual functioning in childhood endocrinopathies and related cytogenetic disorders. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, Pennsylvania, W. B. Saunders, 1969, Chapter 21, p. 1004-1014.

The relation of intelligence to endocrine disorders varies; hypothyroidism and Klinefelter's syndrome are examples of impaired intelligence while adrenogenital syndrome and idiopathic sexual precocity are probably associated with high intelligence. Further, the variation within a syndrome is great, as

- 2663 PADDOCK, NANCY. A comparison of intellectual development of children with spina bifida with their normal siblings. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 107-108.

The problem solving ability of 54 children with spina bifida was compared with that of 58 of their normal siblings. There were no significant differences between the 4 1/2- to 8 1/2-year-old groups, marked differences on all tasks between the 8 1/2- to 12 1/2-year-old groups, and differences on the motor encoding test between the 12 1/2 to 18-year-old groups. All Ss were administered the Motor Encoding subtest of the Illinois Test of Psycholinguistic Abilities, the Similarities subtest of the Wechsler Intelligence Scale for Children, and a Picture-Object test. No significant between group differences were found for the Picture-Object test for any age level. Children in both groups chose objects at a higher level than they were able to verbalize. Children with spina bifida differed from their normal siblings in their problem-solving approaches. On the Similarities subtest, they exhibited inferior performance until adolescence; however, their motoric responses were comparable to those of their siblings at an early age, but at older age levels these response abilities did not advance as far as those of their siblings. These findings confirm Piaget's order of developmental stages and suggest that the time when any stage emerges may vary considerably from child to child. In addition to concrete and abstract conceptualization levels, many Ss (normal and spina bifida) demonstrated primitive styles of thinking. (No refs.)

J. K. Wyatt.

2664 BADELL-RIBERA, ANGELES, SHULMAN, KENNETH, & PADDOCK, NANCY. The relationship of non-progressive hydrocephalus to intellectual functioning in children with spina bifida cystica. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 23-31.

A retrospective analysis of the neurological and psychological status of 75 patients (CAs 5-21 yrs) with varying degrees of spinal cord dysfunction secondary to spina bifida cystica revealed that mean full scale Wechsler Intelligence Scale for Children (WISC) scores and frequency of non-progressive hydrocephalus varied with the degree of disability. Ss with the greatest physical disability had the lowest WISC scores (mean 81; range 45-121) and the highest incidence of hydrocephalus. Among Ss with the greatest physical disability, a subgroup with a history of hydrocephalus had significantly lower verbal, performance, and full scale WISC scores than a subgroup without hydrocephalus. The mean performance IQs of Ss in the hydrocephalic subgroup were significantly lower than their mean verbal IQs. The psychological test findings in hydrocephalic Ss showed characteristic signs of brain damage. Non-progressive or arrested hydrocephalus in the child with spina bifida appears to be a disabling condition which reduces rehabilitation potential. Effective neurosurgery should be performed at an early age in children with spina bifida cystica to increase the survival rate and to improve the quality of the children who survive. (10 refs.) - J. K. Wyatt.

2665 WILNER, ELLIOT, CANNON, JOYCE, & BRODY, JACOB A. Measles, minor neurological signs and intelligence. *Developmental Medicine and Child Neurology*, 11(4):449-454, 1969.

Neurologic examinations and intelligence and achievement tests of 132 school children aged 9-13 years, selected only on the basis of the age at which they had had measles, did not reveal any significant relationship between early infection with measles and signs of minimal brain damage. The mean IQ score and level of reading achievement for the children who had minor neurologic signs were significantly lower than they were for the children who were neurologically normal.

Mixed laterality, or incomplete cerebral dominance, was also found to be significantly more frequent among children with minor neurologic signs. (13 refs.) - *Journal summary*.

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2666 KAELEBER, CHARLES T., & *PUGH, THOMAS F. Influence of intrauterine relations on the intelligence of twins. *New England Journal of Medicine*, 280(19):1030-1034, 1969.

An association of a higher IQ for the member of twin sets with the heavier birth weight, already described for highly selected groups of twins, is reported for a numerically larger and more representative sample. For the 374 twin sets studied, a higher IQ for the heavier twin is found for like-sex pairs with large birth-weight differences (300 gm or more). When information on zygosity is considered, an association is shown for monozygous sets with large birth-weight differences, but none is demonstrated for like-sex dizygous pairs. Unequal intrauterine relations, perhaps of a circulatory nature, are proposed as an explanation for the IQ differences observed among the monozygous sets. (8 refs.) - *Journal abstract*.

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665 Huntington Avenue
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2667 LAURENCE, K. M. Brain damage in hydrocephalic patients. *Proceedings of the Royal Society of Medicine*, 60(12):1265-1266, 1967.

Of 70 hydrocephalic patients who had psychometric testing 6 years earlier and were retested, the majority had maintained reasonable mental development. The mean IQ of the group was unchanged; 40 persons remained the same, 15 persons improved their scores, and 10 hydrocephalics, including all with initial scores below 30 IQ, had deteriorated. A low correlation was found between severity of the hydrocephalus and the degree of mental handicap. However, several persons who had a normal IQ had severe hydrocephalus (cerebral

mantle < 1 cm). Also, severely affected persons exhibited more evidence of brain damage during psychometric and neurological examination. Although cortical thinning undoubtedly causes brain damage as evidenced by the relationship of thinning to IQ, the many severe cases with normal IQ and few signs of

brain damage indicate that the pathology which causes the hydrocephalus may independently be responsible for the brain damage. (3 refs.) - W. J. Klein.

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DEVELOPMENTAL ASPECTS - SOCIAL

2668 ALBIZU-MIRANDA, CARLOS, & STANTON, HOWARD R. The socioeconomic of mental retardation. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 23, p. 183-191.

When a difficult test like the Wechsler Intelligence Scale for Adults was used as a psychometric standard for the measurement of functioning intelligence, almost 1/3 (31.6%) of the adult population of Puerto Rico was classified as MR. Of the 24.6% of the males estimated as MR, 3.2% were estimated to have actual intellectual deficits which presented a problem to society in that they were unable to cope with their environment. The prevalence of MR among lower class women was considerably higher than among lower class men--a factor which appeared to be due to the restricted environment of the women. When the income level of 20% of the community was used as a standard, the majority of the MRs were financially successful. MR was less handicapping in peasant and plantation communities and the rate of failure steadily increased with the complexity of society. Among MR males, 37.4% of those with 4 years or less of education and about 30% of those with more than 4 years of education were unsuccessful. These data suggest that MRs are penalized in more complex societies and demonstrate the need for rehabilitation training programs which will equip MRs to compete with their normal peers. (No refs.) - J. K. Wyatt.

2669 DINGMAN, HARVEY F. Adjustment of the mentally retarded in the community today. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 25, p. 199-202.

The concept of disengagement may prove to be a useful mode for investigating the problem of social adjustment of MRs in the community.

To progress in society, an individual must assume a number of roles. The degree to which he is able to assume a sufficient number of successful roles is the degree to which he is engaged in society. The processes of engagement and the limitations of the roles that MRs can and will assume have been described, and the value of the institution and its social services in providing intervention programs for individuals leaving institutions has been established. To make the outcome of treatment totally effective, stereotyped descriptions of MRs need to be changed, their success in acquiring new roles needs to be evaluated, and techniques for analyzing the factors involved in the successful acquisition of specific social roles need to be developed as well as evaluative techniques for determining community needs for planning and program expansion. (12 refs.) - J. K. Wyatt.

2670 KINGSLEY, RONALD F. Activities and interests of inner city early adolescent educable mentally retarded boys. *Education and Training of the Mentally Retarded*, 3(2):57-62, 1968.

Analysis of activities and personal interests of inner city EMR adolescent boys shows that there is participation and desire for group and active-type activities rather than those requiring passive activity. The one exception was in current indoor activities which were mainly of the passive, solitary type. To measure these interests the Interest-Activities Inventory was constructed by asking 73 boys (CA range 7 to 15 yrs) who were enrolled in regular and special educational classes and who lived in urban, rural, and suburban residential areas to list activities and interests of the past year. The 269 items were placed into 7 categories (indoor, social, hobbies, outdoor, art-music, household chores, and sports). This inventory was presented orally to 149 EMR students (CA range 11 to 15 yrs) approximately 1/2 of

whom were Negro. Results indicated that this selected population preferred participation and group activities. Such data should aid in the development of a meaningful instructional skill program by finding appropriate activities to meet the interests of this population. (18 refs.) - B. Bradley.

Kent State University
Kent, Ohio 44240

2671 RUCKER, CHAUNCEY N., HOWE, CLIFFORD E., & SNIDER, BILL. The participation of retarded children in junior high academic and nonacademic regular classes. *Exceptional Children*, 35(8):617-623, 1969.

The sociometric status of 23 retarded students (mean IQ 71; mean CA 14 yrs 9 mos) integrated into classes with normal children was examined by using the Ohio Social Acceptance Scale. This test consists of 6 descriptive passages ranging from high acceptance to high rejection; the Ss are asked to indicate which of their peers fit the various descriptions. This process yields a social position score. The scores of the MRs were significantly lower ($p < .01$) than the normals in both academic (science) and non-academic (physical education) classes. In both classes, the retarded Ss overestimated their own levels of acceptance. The more popular children in the special classes tended to be more accepted by the normal children. These findings question the efficacy of classroom integration of retarded and normal children and point to the need for the further research. (13 refs.) - J. M. Gardner.

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2672 SHELLHAAS, MAX D. The effects of small group interaction on sociometric choices of day campers. *American Journal of Mental Deficiency*, 74(2):259-263, 1969.

This report demonstrates that small group interaction in a day camp setting increased sociometric choices within the small groups. Cross-choices between institutional retarded campers and non-retarded community campers were also facilitated by the small group interaction. The findings are in support of Homans' theory of a quantitative relationship between interaction and sentiment under generally favorable situational conditions. (14 refs.) - *Journal abstract*.

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2673 TALKINGTON, LARRY W., & HALL, SYLVIA M. Hearing impairment and aggressiveness in mentally retarded. *Perceptual and Motor Skills*, 28(1):303-306, 1969.

Analysis of types of aggressiveness in relation to hearing impairment in MRs shows that the hearing-impaired group scored higher on the self-destructive behavior variable and lower on the attacking peers variable than did the normal hearing group. The hearing-impaired Ss included 56 residents from the Boulder River School and Hospital who had a bilateral hearing loss in excess of 50 decibels as measured by pure tone audiometry at the speech frequencies of 500, 1,000, and 2,000; they were matched for sex, CA, date of admission, and level of MR with a normal hearing control group. These 2 groups were analyzed on 10 aggressive behavioral variables as measured by the 1967 WICHE Population Census Standard Form--a ward personnel rating scale. (11 refs.) - B. Bradley.

Pacific State Hospital
Pomona, California 91766

2674 VOGEL, WILLIAM, KUN, KAROLY J., MESHORER, EDWARD, BROVERMAN, DONALD M., & KLAIBER, EDWARD L. The behavioral significance of EEG abnormality in mental defectives. *American Journal of Mental Deficiency*, 74(1):62-68, 1969.

A group of 45 retardates with abnormal EEGs were matched for age and diagnosis with a group of 45 retardates with normal EEGs. The groups were compared regarding test intelligence, school and occupational performance, personal skills, social behaviors, and psychiatric status for 3 separate periods: year of admission, year of EEG examination, and current year. EEG abnormality is associated with deficits on intelligence test performance, but not with deficits in broader categories of adjustment. However, mean frequency of the EEG is associated with behavioral adjustment as reflected in personal skills and social behaviors, as well as intelligence test and classroom performance. (12 refs.) - *Journal abstract*.

2675 BARRETT, BEATRICE H. Behavioral individuality in four cultural-familially retarded brothers. *Behavior Research and Therapy*, 7(1):79-91, 1969.

The responses of 4 MR brothers (CAs 8 1/2 to 14 yrs; IQs 48-72) to the identical automated

environment were compared. Their education, physical condition, speech, and hearing were basically similar, and all were independently diagnosed cultural-familial retardates. The experimental task consisted of pressing the left of 2 plungers when the left of 2 lights was on. Reinforcement was delivered for every tenth response under this condition, and not for any other combination of plunger presses and lights. During the first hour, one brother performed at the level of a normal adult, discriminating between the two lights and differentiating the use of the plunger. Two brothers failed to either discriminate the light or differentiate between the plungers, while the fourth brother performed at an intermediate level. These striking differences in performance were maintained over 7 experimental sessions. Because of such differences in behavior, the assumption of homogeneity among cultural-familial retardates is questioned. (15 refs.) J. M. Gardner.

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Waverley, Massachusetts 02178

2676 JOHNSON, RONALD C., & ABELSON, ROBERT B. The behavioral competence of mongoloid and non-mongoloid retardates. *American Journal of Mental Deficiency*, 73(6):856-857, 1969.

Comparison of 2,606 mongoloid Ss (mean CA 21.8 yrs; mean IQ 28.6) with 20,605 non-mongoloids (mean CA 24.5 yrs; mean IQ 32) revealed that the mongoloids display greater social competence than non-mongoloids. Behavior which the mongoloids exhibited significantly ($p=.01$, test of significance of difference between percents) more frequently than the non-mongoloids were: dress themselves, understand others, brush their teeth, feed themselves, use the toilet independently, never or infrequently wet the bed, and are considered candidates for ward helpers. Communicating understandably to others occurred significantly less frequently among the mongoloids. The competence of both groups when considered as candidates for work projects, when on a work reward system, and in terms of grooming behaviors was similar. (3 refs.) - A. Huffer.

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2677 MULHERN, THOMAS, & *BAUMEISTER, ALFRED A. An experimental attempt to reduce stereotypy by reinforcement procedures. *American Journal of Mental Deficiency*, 74(1): 69-74, 1969.

Stereotyped behavior is defined as a constant repetition of responses that have no apparent adaptive consequences for the organism. The research reported here attempted to reduce this behavior in 2 SMR, brain-damaged Ss. Two experiments were conducted in which reinforcement was made contingent upon sitting still. Discriminative stimuli were provided to inform the Ss that no reinforcement would be forthcoming while they engaged in stereotyped behavior. The overall effect of these treatments was a reliable reduction in the rate of activity for each S. (9 refs.) *Journal abstract.*

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2678 HALPERN, ANDREW S., & EQUINOZZI, ARTHUR M. Verbal expressivity as an index of adaptive behavior. *American Journal of Mental Deficiency*, 74(2):180-186, 1969.

Verbal expressivity and intelligence were evaluated as differential predictors of level of adaptive behavior for a sample of MR Ss within an institutional setting. Support was found for the hypothesis that verbal expressivity and intelligence are relatively independent of each other and that they do predict different aspects of adaptive behavior. (15 refs.) - *Journal abstract.*

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2679 DILLER, LEONARD, PADDOCK, NANCY, BADELL-RIBERA, ANGELES, & SWINYARD, CHESTER A. Verbal behavior in children with spina bifida. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 100-106.

Ratings of the verbal behavior of 32 children with spina bifida associated with myelomeningocele and hydrocephalus disclosed that younger, more severely handicapped Ss had a clearly aberrant pattern of verbal behavior (dysverbal behavior) which gradually became normal during adolescence. Verbal behavior was rated by observers trained in the medical

and paramedical sciences as either hyperverbal, not hyperverbal, or equivocal. Fifty percent of the Ss had normal verbal behavior, 28% were rated hyperverbal, and 22% were rated equivocal. Hyperverbal Ss differed from normal verbal Ss in that they made significantly ($p < .05$) more irrelevant answers or guesses and fewer ($p < .05$) admissions of ignorance when in situations beyond their competence. They also had significantly ($p < .05$) lower scores on the performance scale of the Wechsler Intelligence Scale for children, and they showed significantly ($p < .05$) more distortions in perception on the Rorschach test. Hyperverbal Ss had a significant ($p < .05$) history of hydrocephalus, were significantly ($p < .002$) more severely disabled, and had poorer performance in daily activities ($p < .05$) and ambulation ($p < .02$). The parents of hyperverbal Ss were significantly ($p < .05$) less likely to have a positive attitude toward them, did not understand their disability ($p < .05$), and failed to follow physicians' instructions ($p < .02$). The dysverbal behavior pattern of the hyperverbal Ss was not conspicuous in stressful situations and was characterized by irrelevant talk. Although this pattern may be influenced by neurologic impairment, it seems to be more related to severity of physical limitations, dependency, excessive time spent with adults, and emotional problems. (16 refs.) - J. K. Wyatt.

2680 WILLIAMS, CYRIL E. Psychiatric implications of severe visual defects for the child and for the parents. In: Gardiner, Peter, MacKeith, Ronald, & Smith, Vernon, eds. *Aspects of Developmental and Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p.110-119.

One of the greatest hazards to a blind child is maladjustment, but many mechanisms may cause this to occur. Statistics indicate that 36% of blind children and 32% of the partially sighted are maladjusted, and approximately 1/3 of all blind children are considered unsuitable for education at school. Abnormal mannerisms of sightless children include manipulation of the eyeballs, symmetrical flapping of the hands, body swaying, and

facial grimacing. Most autistic blind children are a result of inadequate stimulation during childhood, while blind children using excessive speech are attempting to control their environment. The SMR blind child is best cared for in the home because there are few institutions which offer sufficient stimulation and meaningful playthings. Many developmental deviations of blind children are due to lack of interaction between mother and child. This interaction can be improved if the mother makes frequent use of sound and directs the head or body of the infant toward these sounds. Also important is the mother's need to gain information about the care and development of a blind child in order to prevent parental distress which leads to rejection of the child. Public services which provide techniques of handling and guiding a blind child must be extended to the parents. (15 refs.) - V. G. Votano.

2681 DesLAURIERS, AUSTIN M., & CARLSON, CAROLE F. *Your Child Is Asleep: Early Infantile Autism--Etiology, Treatment, Parental Influences*. Homewood, Illinois, Dorsey, 1969, 402 p. \$11.35.

The syndrome of early infantile autism is characterized by grossly retarded learning ability and an almost complete inability to relate to other human beings. A neurophysiological model suggests that autism may be the result of an inborn condition of "imbalance" in which the ascendances of the classical arousal system and the limbic mid-brain arousal system are reversed. This reversal imposes a condition of sensory deprivation on both hypoactive, hyposensitive and hyperactive, hypersensitive autistic children. Research hypotheses on which the treatment of 5 autistic children (CA 2 to 6 yrs) was based were that a highly charged affective climate of human interaction, which included a wide amount and variety of sensory messages communicated through stimulation, would transcend the affective barrier and permit the development of appropriate affective reactions and intelligent, adaptive behavior and that parents can be taught to be effective and normal parents to an autistic child. The psychotherapeutic process was designed to overcome the affective imbalance, extend the child's world, help the child discover his

own body-self and identity, and overcome negativism. Parents were used as co-therapists to help them learn to behave as normal parents and to provide treatment continuity. Treatment periods ranged from 9 months to 2 years. Post-treatment follow-up data showed that all children continued to be affectively involved with other people and that progress has been mainly in speech and social competence. However, progress has not been as rapid as under the high affective impact therapeutic situation, progress in self-sufficiency and affective social adaptability has been slow, and parents have experienced periods of despondency which have interfered with their responses to their children. This book would be of interest to psychiatrists, psychologists, educators, special educators, speech therapists, and parents of autistic children. (119-item bibliog.) - J. K. Wyatt.

CONTENTS: The Language of Childhood; The Circle of Silence; The Communication Barrier; The "Reizchutz" (Stimulus Barrier); The Sleeping Child; A Child of Reality or a Child of Fantasy; How Can Kathy Wake Up? Designing Kathy's Awakening; Kathy, Her Friends, and Their Families; Wake Up Kathy! "O Mother I Am Lost"; Testing of Autistic Children; "And Their Brand-New Eyes Opened to a Brand New Morning!" (Peggy); A Brave New Family; And She Walked; The Child's World of Communication; The Child Is Not Dead ... She is Sleeping.

2682 CLANCY, HELEN, DUGDALE, ALAN, & RENDLE-SHORT, JOHN. The diagnosis of infantile autism. *Developmental Medicine and Child Neurology*, 11(4):432-442, 1969.

The results are presented of the first part of a study designed to clarify the diagnosis of infantile autism. The 9-point scale devised by Creak and others in 1961 was used as the source of reference for data collection. The symptoms of a group of children clinically diagnosed as having infantile autism were analyzed. Control groups of normal children, children diagnosed as rubella deaf, as cerebral palsied and as MR, were compared with the autistic test group. By using the method of numerical taxonomy, 14 major manifestations of infantile autism were isolated. The following criteria are established for the diagnosis of this condition: no one item of the 14 major manifestations is significant in isolation; the diagnostic value of the scale is in the grouping of the symptoms; and a child must show a minimum of 7 items before the diagnosis of infantile autism can be considered. As originally specified by Kanner, the child's symptoms must begin in the first 3 years of life. (13 refs.) - *Journal summary*.

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DEVELOPMENTAL ASPECTS - PSYCHODIAGNOSTICS

2683 ALZOBIAIE, ABDUL JALIL, *METFESSEL, NEWTON S., & MICHAEL, WILLIAM B. Alternative approaches to assessing the intellectual abilities of youth from a culture of poverty. *Educational and Psychological Measurement*, 28(2):449-455, 1968.

Because standard assessment methods are not generally suited for disadvantaged children, there is a need to explore alternative approaches. Guilford's work on creativity provides one possible source. The relation of grade-point average to the Lorge-Thorndike Intelligence Test, the Cattell Culture-Fair Intelligence Test, a reading achievement test

and 3 of Guilford's tests of creativity was evaluated for 122 tenth-grade black students (55 boys, 67 girls). While the reading measure had the highest correlation with grade-point average ($r = .56$), the composite scores for the Guilford tests and the Lorge-Thorndike Intelligence Test were similar (.48 and .46, respectively), and both were higher than the Cattell test. While further research is needed, this study shows the potential of the Guilford approach. (10 refs.) - J. M. Gardner.

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Los Angeles, California

2684 HUTTON, JERRY B. Practice effects on intelligence and school readiness tests for preschool children. *Training School Bulletin*, 65(4):130-134, 1969.

The effect of practice on psychological test performance was studied in 68 children (33 boys, 35 girls; CA range 5 yrs 7 mos to 6 yrs 10 mos; 55 black, 13 white), enrolled in a summer head start program. None had previous experience with psychological tests. The Slosson Intelligence Test (SIT) and the Sprigle School Readiness Screening Test (SSRST) were administered in random order to the Ss, after which the Scholastic Test of Academic Readiness (STAR) was given. Ss taking the SSRST first scored higher than those who took it second; the Ss taking the SIT first scored lower than those who took it second. There were no differences on the STAR. Thus, practice in taking tests had positive or negative effects depending upon the nature of the test. (13 refs.) - J. M. Gardner.

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Richmond, Texas 77469

2685 FULLER, GERALD B., & HAWKINS, WILLIAM F. Differentiation of organic from nonorganic retarded children. *American Journal of Mental Deficiency*, 74(1):104-110, 1969.

A new scoring system was devised for use with the Minnesota Percepto-Diagnostic Test (MPD). When loss of Gestalt and separations in MPD drawings are used as 2 new predictors for brain damage (BD), the Lykken-Rose (1963) disjunctive method of cross-validation yields an 86% hit rate among institutionalized MR 10- to 16-year olds diagnosed as BD and an 85% hit rate among a similar group of retardates though diagnosed not BD. The average 85% correct identifications is considerably above other predictors used to differentiate BD from non-BD retardates. (7 refs.) - *Journal abstract*.

Central Michigan University
Mount Pleasant Michigan 48858

2686 TRITES, RONALD L., & MATTHEWS, CHARLES G. Psychologic test findings in the F-form of acro-pecto-vertebral dysplasia: The F-syndrome. In: Bergsma, Daniel, ed. *Birth Defects: Original Article Series, Part III*, 5(3):64-67, 1969.

A battery of psychological tests was given to 8 members of the "F" family, 5 of whom were

affected with a syndrome of acro-pecto-vertebral dysplasia. Verbal and performance IQ scores and academic achievement scores were lower in the affected group. Sensory discrimination and nonverbal abstraction test results were normal in both groups. The affected patients achieved lower scores on motor proficiency tests, but this may be explained, in part, by their hand anomalies. Exceptions to group trends were noted in both groups. (13 refs.) - E. L. Rowan.

2687 NIHIRA, KAZUO. Factorial dimensions of adaptive behavior in mentally retarded children and adolescents. *American Journal of Mental Deficiency*, 74(1):130-141, 1969.

Objective descriptions of adaptive behavior of 313 institutionalized MR children and adolescents were obtained by means of a newly developed behavior rating scale. Factor analyses of the scale scores delineated 3 major dimensions--Personal Independence, Social Maladaptation, and Personal Maladaptation. Comparison of the results between 4 different age groups reveals that the obtained factor structure is relatively stable across a wide span of age ranges, from pre-adolescent through adulthood. Implications of the results are discussed in terms of adaptive mechanisms commonly used by retardates in coping with their environments. (24 refs.) - *Journal abstract*.

Parsons State Hospital and Training Center
Parsons, Kansas 67357

2688 BROWN, DUANE, & JONES, ELLA. Using the Bell Adjustment Inventory with the mentally retarded. *Rehabilitation Counseling Bulletin*, 13(3):288-294, 1970.

The Bell Adjustment Inventory (BAI) was given to a group of 150 EMR students. When comparisons were made with a normal distribution it was found that the retarded group had significantly more members in the low-adjustment categories on the home adjustment, health adjustment, hostility, emotionality, and submissiveness subtests. Boys had significantly lower scores on the masculinity subtest, but scores for retarded girls were distributed normally on the femininity subtests. Recommendations are made for use of the BAI in counseling the MR. (18 refs.) - *Journal abstract*.

2689 DIERKS, DARRELL & CUSHNA, BRUCE. Sex differences in the Bender Gestalt performance of children. *Perceptual and Motor Skills*, 28(1):19-22, 1969.

The open question of sex differences in the diagnostic use of the Developmental Bender Scoring System for Young Children remains a troubling issue. These differences have been often obscured by the failure to designate clear diagnostic categories or to control for maturational effect. The present study accounts for each of the above variables and their interaction through multivariate design. The presence of a significant sex difference among these 478 children from a clinic population demonstrates the need for further exploration of sex differences in visual motor performance. (8 refs.) - *Journal abstract*.

University of Iowa
Iowa City, Iowa 52240

2690 ALLEN, ROBERT M. The developmental test of visual perception and the Bender Gestalt Test achievement of educable mental retardate. *Training School Bulletin*, 66(2): 80-85, 1969.

The performance of 36 EMRs (mean CA 161 mos; mean IQ 57) on the Bender-Gestalt (BG) and the Frostig Developmental Test of Visual Perception (DTVP) was measured, and the relation between the tests examined. The retardates scored significantly below normal on the BG and all sub-scales of the DTVP including the total score. BG scores were significantly correlated with the total and sub-scaled DTVP scores, indicating that poor perception (as measured by the DTVP) and poor motor coordination (as measured by the BG) go together. BG scores, but not DTVP scores, correlated significantly with intellectual level (as measured by the Peabody Picture Vocabulary test). When the Ss were divided into high and low perceivers according to their DTVP scores, it was found that high perceivers had significantly better ($p < .01$) BG scores. In planning for the educable retardate, it is important to consider visual perceptual and motor abilities. (13 refs.)
J. M. Gardner.

University of Miami
Coral Gables, Florida 33146

2691 ALLEY, GORDON R. Comparative constructional praxis performance of organically impaired and cultural-familial mental retardates. *American Journal of Mental Deficiency*, 74(2):279-282, 1969.

Two groups of MR children were compared on a constructional praxis task (Benton Visual Retention Test). One group was diagnosed as organically impaired (N=20) and another group was classified as cultural-familial retarded (N=20). The latter group obtained significantly higher absolute scores on 10 of 18 variables investigated. It was concluded that, as a constructional praxis task, the Benton Visual Retention Test is useful in the differential diagnosis of organic impairment in MR children. (9 refs.) - *Journal abstract*.

University Hospitals
Iowa City, Iowa 52240

2692 BELLAMY, EDWARD, & DALY, WILLIAM C. The height of figure drawings related to IQ, sex, and CA in mental retardates. *Journal of Clinical Psychology*, 25(2):206-207, 1969.

The relation between height of human figure drawings produced by 129 institutionalized MR, (IQ range 35-80; CA range 13 to 20 yrs; 73 males, 56 females) and IQ, sex, and CA was investigated. The mean heights of the drawings for the 3 IQ groups were 115 mm for those in the 35-50 IQ group (N=38); 121 mm for those in the 51-65 IQ group (N=59), and 150 mm for those in the 66-80 IQ group (N=32). The *t* tests revealed that the size of the drawings by the 66-80 IQ group exceeded the other 2 groups, who did not differ. The overall effect was significant at the .05 level (*F* test). No significant differences were found between drawings of males and females, nor was there any significant relation between CA and height of drawing. Both sexes (41% females, 26% males) drew the opposite sex first more frequently than normal Ss. (3 refs.) - J. M. Gardner.

A. L. Bowen Children's Center
Harrisburg, Illinois 62947

2693 WANDERER, ZEV WILLIAM. Validity of clinical judgments based on human figure drawings. *Journal of Consulting and Clinical Psychology*, 33(2):143-150, 1969.

Draw-A-Person (DAP) protocols were obtained from 5 diagnostic populations: retardates, homosexuals, psychotics, neurotics, and normals. The groups were matched on age, ethnic status, socioeconomic class, marital status,

and other relevant variables. Twenty individuals who were reputed experts in the diagnostic use of the DAP were asked to classify the protocols. While the overall results indicated that the experts could identify the populations better than chance, this was true only for the retardates and no other group. The retardates were identified in almost 100% of the cases. (31 refs.) - J. M. Gardner.

Center for Behavior Therapy
Beverly Hills, California 90210

- 2694 DILORENZO, LOUIS T., & BRADY, JAMES J.
Use of the Peabody Picture Vocabulary Test with preschool children. *Training School Bulletin*, 65(4):117-121, 1969.

While the Peabody Picture Vocabulary Test (PPVT) provides an easily administered and scored measure of intellectual functioning, there are numerous limitations to the use of the test. First, the use of large age intervals (6 months) often produces large IQ differences for children separated by only one month of age. Second, it has been found that PPVT-derived IQ scores often deviate from IQ scores derived from established intellectual tests. In a study of 563 3- and 4-year old children, mean differences of more than 8 points were found. A third limitation in the PPVT is the lack of comparable growth in IQ when advancing from one age level to another. A final limitation is the lack of comparable changes on either side of the mean; equal scores will produce different IQ point changes depending on whether they are above or below the mean for the age level. Users of the PPVT should be aware of these limitations. (10 refs.) - J. M. Gardner.

New York State Education Department
Albany, New York 12200

- 2695 SHOTWELL, ANNA M., O'CONNOR, GAIL, GABET, YVONNE, & DINGMAN, HARVEY F. Relation of the Peabody Picture Vocabulary Test IQ to the Stanford-Binet IQ. *American Journal of Mental Deficiency*, 74(1):39-42, 1969.

The Peabody Picture Vocabulary Test (PPVT) has been recommended as a substitute for the Revised Stanford-Binet (S-B LM) for use in assessing mental ability. Correlation analyses indicate that the relation between PPVT and the S-B LM is relatively strong. Regression analysis and a bivariate frequency distribution, however, indicate that it is not possible to predict perfectly S-B IQs from the PPVT IQs. A method for computing S-B IQs

from PPVT IQs is discussed. (6 refs.)
Journal abstract.

Pacific State Hospital
Pomona, California 91766

- 2696 CONGDON, DAVID M. The Vineland and Cain-Levine: A correlational study and program evaluation. *American Journal of Mental Deficiency*, 74(2):231-234, 1969.

The progress of 23 TMR male Ss in a 1-year institutional training program was evaluated by a test-retest procedure with the Cain-Levine Social Competency Scale and Vineland Social Maturity Scale. Initially, the instruments were found to be significantly correlated with each other and MA. Both scales indicated improvement in social competency but the Cain-Levine had the additional advantage of discriminating between areas of progress which corresponded to program emphasis. Initially less competent Ss achieved more over the 1 year than did the initially more competent Ss. (12 refs.) - *Journal abstract.*

Lincoln State School
Lincoln, Illinois 62656

- 2697 SILVERSTEIN, A. B. WISC subtest patterns of retardates. *Psychological Reports*, 23(3):1061-1062, 1969.

The summarized results of 10 published studies which reported Wechsler Intelligence Scale for Children subtest patterns of MRs show that the MRs' greatest strengths lie in the performance factor while their greatest weaknesses lie in the verbal factor. The easiest to the most difficult subtests ranked as follows: Object Assembly, Picture Completion, Block Design, Coding, Similarities, Picture Assembly, Comprehension, Information, Arithmetic, and Vocabulary. Despite differences among the groups, the average correlation among the rankings was .68. (12 refs.)
A. Huffer.

Pacific State Hospital
Pomona, California 91766

2698 BARCLAY, A., FRIEDMAN, ELLEN C., & FIDEL, YILDIZ. A comparative study of WISC and WAIS performances and score patterns among institutionalized retardates. *Journal of Mental Deficiency Research*, 13(2):99-105, 1969.

To compare test performance patterns among MRs, the Wechsler Adult Intelligence Scale (WAIS) was given to 113 institutionalized MRs (CA range 16-35 yrs; IQ range 40-84) and the Wechsler Intelligence Scale for children (WISC) was administered to 63 residents (CA range 9-16 yrs; IQ range 40-84) from the same institution. The sex ratio for the WAIS group was 73 males, 40 females while the WISC group contained 47 males and 16 females. Selection of Ss was done randomly. Results indicate no significant differences in overall test performance although there were some variations in score patterns as 3 of the subtests (Similarities, Arithmetic, and Digit Symbol) showed significant differences. The fact that performance skills were more highly developed than verbal skills is possibly due to the general cognitive style of the retardate. (11 refs.) - B. Bradley.

St. Louis University
221 North Grand Boulevard
St. Louis, Missouri 63103

2699 BASSETT, JOHN E., & GAYTON, WILLIAM F. The use of Doppelt's abbreviated form of the WAIS with mental retardates. *Journal of Clinical Psychology*, 25(3):276-277, 1969.

Correlation of 110 MR adults (CA range 16 yrs 3 mos to 59 yrs 11 mos) Wechsler Adult Intelligence Scale (WAIS) full scale IQs (45 to 78, mean 60) with Doppelt's short form of the WAIS showed that, when the short form is used, a correction factor is not needed. Doppelt's IQ was calculated on the basis of 4 of the 11 subtests. In addition, a corrected factor of 2 was subtracted from each Doppelt IQ. The WAIS full scale IQ correlated .87 (standard error of estimate = 3.28) with the Doppelt IQ and .87 (standard error of estimate = 3.29) with the corrected Doppelt IQ. The variances did not differ significantly from each other ($t=.61$ for full scale IQ and Doppelt IQ; $t=.58$ for full scale IQ and corrected Doppelt IQ; and $t=.023$ for Doppelt IQ and corrected Doppelt IQ). (2 refs.) - A. Huffer.

Pineland Hospital and Training Center
Pownal, Maine 04069

2700 LOVE, HENRY G. I. Validity of the Doppelt Short Form WAIS in a psychiatric population. *British Journal of Social and Clinical Psychology*, 8(2):185-186, 1969.

Data from the administration of the Doppelt Short Form of the Wechsler Adult Intelligence Scale (WAIS) to 51 psychiatric Ss (CA range 17-66 yrs; WAIS IQ range 46-125) including MRs and epileptics indicated that the short form leads to miscalculation of the Standard Form IQ. The Doppelt IQs were obtained by adding an age constant to the multiplicand of 2.5 and the sum of 4 subtest scaled scores. Tabulation of the IQ differences between the Doppelt and the Standard Form IQs revealed that a range of ± 13 IQ points is needed to obtain 65% correct predictions. Full Scale IQ mean ± 15 IQ points gives a 66% prediction; therefore, "guessing Full Scale IQ from the mean and standard deviation of the Standard Form would be more economical and almost as accurate." (11 refs.) - A. Huffer.

Porirua Hospital
Porirua, New Zealand

2701 FULTON, ROBERT T., & REID, MICHAEL J. Bekesy Audiometry with the retarded. *American Journal of Mental Deficiency*, 74(2):223-230, 1969.

Thirty-one Ss representing 3 measured intelligence levels and 3 hearing type classifications were pretrained to Bekesy response techniques via vibrotactile procedures and then auditorily assessed with standard pure-tone and Bekesy techniques. Bekesy variables included: sweep-fixed procedures, attenuation rate, frequency, and interrupted-continuous stimuli. Excursion sizes were determined for Bekesy conditions. (11 refs.) - *Journal abstract.*

Parsons State Hospital and Training Center
Parsons, Kansas 67357

2702 SPRADLIN, JOSEPH E., LLOYD, LYLE L., HOM, GEORGE L., & REID, MICHAEL. Establishing tone control and evaluating the hearing of severely retarded children. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 22, p.170-180.

Operant techniques were successfully used to evaluate the hearing of 41 SMR children. If

the expected response to a warble tone (70 db, 500 cps) and a light was not established within 10 or 12 aided trials, a shaping procedure was used; under these conditions, control was usually established rapidly. Tone control was then established by a 5-step light-fading process. Intensity and frequency generalization were instituted and usually accomplished without difficulty. When sound field screening was completed, the child was trained to wear earphones so that a bilateral test could

be obtained. This training proved to be more difficult with social reinforcement than with food. Of the 41 SMR children who participated in operant conditioning audiometry, tone control and bilateral hearing evaluations were obtained on 26. Of these, 15 had essentially normal hearing, 2 had unilateral hearing losses, and 9 had hearing losses in both ears. From 7 to 50 sessions were required to obtain hearing evaluations. (8 refs.) - J. K. Wyatt.

TREATMENT AND TRAINING ASPECTS - EDUCATIONAL

2703 BIJOU, SIDNEY W. Research on the academic education of the retarded. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation*. Springfield, Illinois, Charles C. Thomas, Chapter 19, 1968, p. 143-149.

The viewpoint of the educational technology which emerges from experimental analysis of behavior is that failures in academic learning are due to failures of the motivational system and/or inadequate programmed material. In an experimental academic project which applied experimental analysis of behavior principles to the academic education of MR children (mean CA 10 yrs 2 mos; mean IQ 65), programs and practices were developed and evaluated by establishing a baseline performance for each child or by using experimental operations. A waiting room was used to help calm the children before they entered the classroom and to emphasize the schoolwork function of the classroom. The reinforcement system included the use of a "timeout" room as a quiet place for children whose behavior problems interfered with learning and the reinforcement of all desirable responses with social reinforcers or tokens. Programs in reading, writing, and arithmetic which are based on programmed instruction principles and on analysis of the performance of each child have been devised. The research orientation of this project emphasizes the performance of the individual child rather than gross differences in performance between classes of children who have been exposed to different educational programs. (9 refs.) - J. K. Wyatt.

2704 BROWN, LOUIS F. The special class: Some aspects for special educators to ponder. *Education and Training of the Mentally Retarded*, 3(1):11-16, 1968.

Special education classes for EMRs are being labeled ineffective; however, rather than the underlying assumptions for special education being false, the actual class environments do not adhere to these assumptions. The assumption that a low teacher-pupil ratio aids instruction is partly followed in that class size is limited to 15, but it is unknown if instruction is individual. The principle of specially trained teachers is unreal, for 18 states have no special requirements for teachers of MR, only 11 states require student teaching of MR, and in Iowa 37% of teachers have a Bachelor of Arts degree or less. The assumption of close supervision by directors is not upheld, for 28% of their time is for supervising and 7% for in-service education activities. It is assumed that MRs are with the same teacher longer, but lower socioeconomic families (who generate the most EMR) relocate often and teacher turnover is high. The assumed carefully structured continuous curriculum is rare; research on its effectiveness is needed. It is assumed that the special class is more homogeneous, but IQs vary by 35 points; there are differences in age, coordination, achievement, emotional and social age; and the lower IQ indicates brain damage. It is assumed that there are more opportunities for leadership roles in the special classes; however, this is not unique to the special class, for teachers

could arrange these opportunities in a normal class. Conclusions are: ineffectiveness of special classes has not been demonstrated; since few classes fulfill the above assumptions, it is unknown how effective special education could be; future research should separate assumed from actual class conditions; and special educators should try to implement the assumed conditions before condemning the system. (24 refs.) - M. Plessinger.

University of Iowa
Iowa City, Iowa

- 2705 HETZNER, DONALD R. The borderline student as a member of a separate school group. *Journal of Experimental Education*, 37(2):48-52, 1968.

Grades 7-12 need prevocational classes for 17% to 20% of the students who have IQs 75-90 or a severe reading disability. An existing program sought to determine if prevocational classes were justified. Of 168 students in the program, 98 were admitted because they had IQs between 75-90, and 70 were enrolled because they were reading at least 2 grades below their age. Recent intelligence test scores (Otis Beta for the prevocational and academic classes; Stanford-Binet or Wechsler Intelligence Scale for Children for the special education classes) showed that the mean IQs of those in the prevocational classes were significantly different from those in special education classes and those in academic classes. Students entering prevocational classes had less absenteeism, completed their homework more often, exhibited fewer behavior problems, and had fewer drop-outs than the other programs. Since there is a school population who needs prevocational classes, schools must recognize the need and design curricula tailored for those slow learners who do not belong in either special education or academic classes. (6 refs.) - M. Plessinger.

Medina School System
Medina, New York

- 2706 LANCE, WAYNE D. School programs for the trainable mentally retarded. *Education and Training of the Mentally Retarded*, 3(1):3-9, 1968.

With the increase of TMR classes, attention must be given to curriculum development,

teacher training, teaching methods, evaluation, and school organization. Clear goals of TMR curriculum must be established and the emphasis shifted from academic achievement. Understanding the characteristics (intellectual, social, motor, physical, and adult behavior) of the TMR is a prerequisite to appropriate curriculum development. Prospective teachers should be carefully screened, and their training should center on: cognitive, perceptive, and sensorimotor development; research and evaluation techniques; language development; leisure-time concepts; vocational education; parental counseling; and the teacher's role as a stimulus to elicit response. Practice teaching in TMR classes is a must. Teachers must become aware of tests and how to use them to evaluate the TMR's physical, social, and self-help competencies and to develop, on the basis of test results, a clinical teaching approach. TMR program reorganization should include: education beginning at 4 years old; year round schools; work experience; preparation for semi-independent community living; and freedom for teacher innovation, team teaching, aides, and cooperation with specialists. (47 refs.) - M. Plessinger.

University of Oregon
Eugene, Oregon 97403

- 2707 STARK, EDWARD S., ed. *Special Education: A Curriculum Guide*. Springfield, Illinois, Charles Thomas, 1969, 256 p.

This concrete and comprehensive guide and course of study includes curricula for MRs, brain injured, and the severely physically disabled. For each unit, the steps required for its completion are described, teacher activities are outlined, and a list of teaching materials is included. Curriculum patterns in the field of MR are broken down into curricula for TMRs, pre-primary and primary EMRs, intermediate EMRs, junior high school EMRs, a work-study program for EMRs, and a sheltered workshop program. This book would be of interest to school administrators, curriculum planners, and special education teachers. (6 refs.) - J. K. Wyatt.

CONTENTS: The Trainable Mentally Retarded-- A Philosophy (La Magna); The Pre-Primary and Primary Special Classes for the Educable Retardate (Farrell); The Intermediate Level Class for the Educable Retardate (Barsky); The Junior High School Special Class for Retarded (Payerle); The Preparation and Placement of the Educable Retardate (Hecht);

A Sheltered Workshop Experience for the Mentally Retarded (Levine); Teaching the Brain-Injured Child--Elementary Level (Zoueff); Teaching the Brain-Injured Child--Junior High School Level (Sinagra); A Curriculum for the Severely Crippled Child (Lombardi); Rounding Out the Curriculum (Stark).

2708 LA MAGNA, SAM A. The trainable mentally retarded--A philosophy. In: Stark, Edward S., ed. *Special Education: A Curriculum Guide*. Springfield, Illinois, Charles C. Thomas, 1969, Chapter 1, p. 5-47.

Classes for TMRs should be centralized so that the basic objectives of education can be achieved. These objectives include the provision of increased homogeneity within a classroom, opportunities to relate to a true peer group and develop social relationships, opportunities for each child to become familiar with the environment, ancillary services, and a global program which is developmental and goal-oriented. The curriculum for TMRs at the Naussau County Special Services School (New York) focuses on the areas of social studies, health, language arts, mathematics, science, sensory concept training, safety habit training, arts and crafts, physical education, and cultural appreciation. Developmental aims and objectives are outlined for each area at primary and intermediate levels. (No refs.) - J. K. Wyatt.

2709 FARRELL, EVELYN. The pre-primary and primary special classes for the educable retardate. In: Stark, Edward S., ed. *Special Education: A Curriculum Guide*. Springfield, Illinois, Charles C. Thomas 1969, Chapter 2, p. 48-85.

Pre-primary and primary special classes for EMRs provide a structured environment in which the children can begin to experience success in school experiences and develop peer relationships. Each child should be provided with the specific learning experiences he requires to meet his individual needs. Visual curriculum areas should not be explored until children with perceptual, speech, and motor control problems receive specific help to overcome them. Curriculum units in the areas of self-help and perception, arithmetic, science, health, safety, language arts, writing, reading, and social studies are described. The steps required to complete each unit are described as well as specific teacher activities and lists of helpful materials. (6 refs.) - J. K. Wyatt.

2710 BARSKY, LEON. The intermediate level class for the educable retardate. In: Stark, Leon S., ed. *Special Education: A Curriculum Guide*. Springfield, Illinois, Charles Thomas, 1969, Chapter 3, p. 86-130.

Special education curricula should include all the school learning experiences in which a child will participate, allow considerable freedom to the teacher and child in regard to the determination of immediate goals and the best methods for their achievement, and provide the teacher with a wide variety of methods and materials. Educational goals for the MR include the development of self, interpersonal and intergroup relations, an understanding of people and things, and the ability to use essential skills. Intermediate curricula are designed for EMRs from 9 to 13 years of age and should include tool subjects (social studies, health and hygiene, language arts, arithmetic, and science) with content based on "areas of living" and may be implemented by using unit, experimental, or traditional subject matter approaches. Each area is broken down into a series of units and teacher activities and pertinent materials are described. (No refs.) - J. K. Wyatt.

2711 PAYERLE, ADELE. The junior high school special class for retarded. In: Stark, Edward S., ed. *Special Education: A Curriculum Guide*. Springfield, Illinois, Charles Thomas, 1969, Chapter 4, p. 131-146.

The junior high school special education class includes MRs from 13 to 16, and its program attempts to bridge the gap between childhood and elementary school and young adulthood. This is a time of anxiety and conflict for many MRs, because their CA and physical size cause people to make demands on them which they cannot fulfill. The classroom situation should be structured so that the children have the freedom to be what they are in a realistic setting so that their behavior can be transferred to the "outside world." Placement in regular nonacademic classes helps MRs to learn to adjust their behavior and increases their social development. Curricula in the areas of arithmetic, language arts, occupational information, and social studies are described. Suggestions for the integration of MR children into regular, physical education, home economics, industrial arts, art, and music classes are included. (No refs.) - J. K. Wyatt.

2712 KOKASKA, CHARLES J. Secondary education for the retarded: A brief historical review. *Education and Training of the Mentally Retarded*, 3(1):17-26, 1968.

Since secondary EMR education is new, well-defined programs are needed. Only 4.6% of the nation's school districts have EMR special education classes. In the 1940s, several large cities set up separate EMR trade classes, for World War II had created a labor demand and MRs proved competent; however, since they went to work at 16, special classes ended there. By 1945, attitudes changed and the high school philosophy was modified to provide EMR education through senior high; public high schools began to prepare persons for work as well as for college. Since economic survival is a basic educational goal, the implementation of this goal shifted to work experience. By the late 1950s, arrangements were being made with businesses for training students. Attention was given to determining the role of the employer in educating students, developing school programs in cooperation with public agencies and private business, and providing a balanced (general vs specific skills) training program. (38 refs.) - M. Plessinger.

Eastern Michigan University
Ypsilanti, Michigan 48197

2713 ZITO, ROBERT J., & BARDON, JACK I.
Achievement motivation among negro adolescents in regular and special education programs. *American Journal of Mental Deficiency*, 74(1):20-26, 1969.

One hundred fifty Negro adolescents (50 EMR in special classes, 50 EMR in regular classes, and 50 Ss of normal intelligence) were compared for achievement motivation and objective goal setting behavior, using thematic apperception pictures, the Wide Range Achievement Test, and a task designed to measure aspiration reality and shift. Results seem to indicate that retarded Negro adolescents are more influenced by success than by failure, retarded Negro adolescents have achievement motivation comparable to normal Ss from the same socioeconomic background, special class experience makes EMR adolescents cautious in setting goals, and special class retardates anticipate failure to achieve goals while regular class retardates anticipate success and, in fact, show greater word recognition achievement. (15 refs.) - *Journal abstract*.

Newark State College
Union, New Jersey 07083

2714 FRIEDLANDER, BERNARD Z. Psychology and the third R in special education. *Education and Training of the Mentally Retarded*, 3(2):80-89, 1968.

Although arithmetic and math are difficult to teach due to abstractions, the MR can learn useful problem solving when instruction is carefully structured to specific principles of psychological development, general principles of thinking, and factors of sight perception. Math is now taught with an operational approach, which stresses working problems of addition, subtraction, multiplication, and division, or from the new math conceptual approach, which emphasizes the teaching of underlying laws such as sets and different number bases. Mental development progresses through 3 stages: the enactive stage when a child relates to everything physically; the intermediate iconic stage when the child accepts pictures in place of actual experience; and the symbolic stage when a number is accepted as representing a quantity. To attain the symbolic stage a child must understand numerosity (objects can be defined quantitatively by a number), conservation (a quantity remains constant even though rearranged), equivalence (different numbers or quantities can equal others although the numbers are not identical), flexible combinations (different combinations of quantities can equal the same quantity), and generality (certain classes have common features despite superficially different appearances). To aid the MR in learning arithmetic, materials must be designed and presented in a perceptually clear manner so as to minimize perceptual distortion problems. (7 refs.) - M. Plessinger.

University of Wisconsin
Madison, Wisconsin 53706

2715 PEACH, WALTER, MONACO, THERESA, BLANTON, RICHARD S., & HURLBURT, GOLDA. Effectiveness of programed instruction related to color discrimination for the trainable retardates. *Central Missouri Synthesis on Mental Retardation*, 1(1):21-26, 1968.

Programed instruction directed to the discrimination of colors with employment of the Edison Responsive Environment showed an improvement in the performance level of 14 of the 26 institutionalized TMR Ss (Stanford-Binet IQs 20-58; CAs 84-264 mos). The Edison Responsive Environment included an electronic typewriter with 6 colored keys, a slide projector, and the "voice of the environment." Each S had a color discrimination program of 5-10 minutes in length for 5 days. Results showed a performance increase for 14 Ss, no

performance change for 9 Ss, and a performance decrease for 3 Ss. Three definite groups were determined: Ss who found this color discrimination task to be too difficult, Ss evidencing an improvement from first to fifth presentation, and Ss receiving high scores on all 5 programs. This type of instruction appeared effective for some of the TMR population studied. (13 refs.) - B. Bradley.

No address

- 2716 KLEINKE, PATRICIA D. Beneficial teaching techniques for the trainable retarded. *Education and Training of the Mentally Retarded*, 3(2):94-96, 1968.

The TMR can learn to care for themselves, be useful to others, be taught to express themselves meaningfully and, by the utilization of beneficial teaching techniques, can grow up relaxed, secure, and happy. A 2-year pilot program in Oregon demonstrated that TMR children can adequately accomplish activities that: develop their social awareness and adjustment; improve their motor skills, coordination, and communication; and develop their self-confidence. Hand plays enhanced their progress and contributed significantly toward better socialization and conversation. Educational art activities provided them with enriching classroom experiences that proved to be of much value and benefit in their everyday living situations. (No refs.) - S. Half.

Cedar Hills Cooperative School
Beaverton, Oregon 97005

- 2717 JORDAN, LAURA J. Effective seatwork for the educable. *Education and Training of the Mentally Retarded*, 3(2):90-93, 1968.

Teachers of EMR classes must be aware of each child's limitations, level of functioning, problems, and capabilities if students are to complete their seatwork effectively and independently. Good seatwork must teach something the child needs to learn, be an activity he can perform independently, and occupy him for the right amount of time with consideration given to his attention span, the difficulty of the work, and the scheduling of the rest of the class. Commercially prepared seatwork material must be evaluated in terms of its value to the child's program. Frequently, the use of nonconsumable seatwork (puzzles, formboards, stamp sets, cards, and stencils) will be a more appropriate and

therapeutic approach. Immediately following completion, all seatwork should be carefully and thoroughly checked. (No refs.) - S. Half.

University of Illinois
Urbana, Illinois 61801

- 2718 CAMENGA, SUSAN J. Instructional materials for use with the mentally retarded. *Education and Training of the Mentally Retarded*, 3(1):27-29, 1968.

Instructional materials to teach MR reading, arithmetic, music, and recreation are listed with a brief description of the goal, intended audience, publication date, price, and source. The materials include books, pictures, films, puppets, blocks, and records which teach community sights, animals, personal health, counting, telling time, singing, rhythm, crafts, and games. (No refs.)

M. Plessinger

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- 2719 GOLD, MARC W. Preworkshop skills for the trainable: A sequential technique. *Education and Training of the Mentally Retarded*, 3(1):31-37, 1968.

TMR can learn useful tasks which require formal training through sequential training of the skills involved in the task. In the sequential technique, only one item is learned at a time, one step is learned before a new one is added, and all tasks are reduced to a series of small steps. The child's current skills and attitudes should be evaluated in terms of the task to be completed. The TMR should progress at his own rate, learn the necessary vocabulary, and continually review earlier steps. Sorting, assembly, and self-direction tasks are described to illustrate the variables to consider when developing a preworkshop curriculum. (No refs.)

M. Plessinger.

No address

- 2720 RADIGK, WERNER. *Arbeitsmittel und Arbeitshilfen im Unterricht der Sonderschulen fuer Lernbehinderte (Teaching Aids and Advice for Special Education Programs for the Mentally Retarded)*. Berlin, West Germany, Carl Marhold Verlagsbuchhandlung, 1968, 130 p. (Price unknown).

Educational principles which apply especially to the teaching of the MR are: use of visual

aids, action teaching, progression in the smallest possible steps, and repetition. Of these, the use of visual aids and repetition are the most important. Methods are described for using various aids (3-dimensional models and games) in the teaching of number concepts, functional reading, and environmental concepts. Once the child has performed tasks under supervision, there should be an opportunity for him to practice individually with the material and with additional material of the same type. The instructor should not overlook any resource and should constantly search out new material. (27-item bibliog.) S. L. Hamersley.

CONTENTS: Theories of Teaching Aids and Materials; Practice of Teaching Aids and Materials; Theory and Practice of Teaching Aids and Materials with Examples of Lesson Plans.

2721 GOEDMAN, M. H., & KOSTER, H. *Hoe moet if verder met dit kind? (How may I make progress with this child?)* Nijkerk, Holland, Uitgeverij G. F. Callenbach, 1969, 155 p. (Price unknown).

The MR child goes through 6 main phases of development: beginning contacts with his surroundings, learning basic reasoning processes, copying the actions of others, trying independent actions, learning how to perform these actions, and becoming independent in his control of these actions. Each of these stages requires a different pedagogical approach in terms of the type and difficulty of tasks given to the child. One important factor at all levels is that expectations of the child's performance should not be placed very high, because the MR are especially subject to frustration and discouragement. At each level, the instructor must encourage the child to respond and act independently, but he must not expect that this behavior be the rule. Progress towards independence will be slow and sometimes regression will be noted. During these periods the supportive role of the instructor is most strongly emphasized. At other times, he may be an observer, seeking out methods to encourage the child's growth in skills or recording progress. (65-item bibliog.) - S. L. Hamersley.

CONTENTS: Who is This child? On the Way to Human Growth and Development; Observe Carefully and Know How to Teach; Continuation of Educational Intent through the Use of Material Aids.

2722 EISENBERG, LEON, & CONNORS, C. KEITH.

The effect of Head Start on developmental processes. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 14, p. 116-122.

An evaluation of the effects of a 6-week Head Start program on selected aspects of cognitive development suggests that a preschool experience and a special form of competent preschool teaching can effect highly significant intellectual gains in preschool children. The Peabody Picture Vocabulary Test (PPVT) and the Draw-A-Person (DAP) were administered to an experimental group (425 Ss) during the first week of a Head Start program, at program termination, and on enrollment in kindergarten and to a control group (378 Ss) on enrollment in kindergarten. Teacher behavior in the classroom was observed by trained observers. Head Start Ss made progressive and substantial gains in oral vocabulary (as measured by the PPVT) at each successive testing. Differences between the Head Start means at the second and third testing and between initial Head Start and control group means were significant ($p < .0001$); the Head Start group continued to be inferior. At the termination of the Head Start program, Head Start Ss had significantly ($p < .01$) higher DAP scores than the control Ss. Teachers rated as warm, flexible, and varied in activity produced significantly ($p < .05$) greater change in the IQs of their students than their opposites. Ss in classes where the teachers spent a high proportion of their time teaching made significantly ($p < .001$) larger IQ gains than Ss in classes where the teachers played or spent considerable time enforcing obedience. (8 refs.) - J. K. Wyatt.

2723 LONGMORE, JOHN B. The education of the visually handicapped child with additional disabilities. In: Gardiner, Peter, MacKeith, Ronald, & Smith, Vernon, eds. *Aspects of Developmental and Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p. 120-121.

Candover Hall, a school for blind children, has 91 children between the ages of 13 and 17, and most of these are educationally subnormal and/or physically handicapped. Of the total group 60 have IQs < 80 , 15 are deaf and blind, 20 have epilepsy, 25 have cerebral palsy, and 16 have severe emotional problems. There is a high staff-pupil ratio (1:2 in the main school and 1:1 in the deaf-blind unit),

and emphasis on teaching independence. Children are able to walk around the grounds or town and eventually go on very distant trips. In order to teach independence, the children participate in activities which include physical education, music, handicrafts, learning braille, scouting, and handling money. Surveys indicate that the "Candover Child" is often absorbed into training centers for sighted handicapped people. (No refs.)

V. G. Votano.

- 2724 REGER, ROGER, SCHROEDER, WENDY, & USCHOLD, KATHIE. *Special Education: Children with Learning Problems*. New York, New York, Oxford University, 1968, 251 p. \$5.00.

This description of an educational program for children whose learning patterns and/or behavior deviate significantly in a negative direction from the average includes critical discussions concerning the nature of special education as well as descriptions of specific curriculum materials, teaching techniques, and methods of evaluating learning problems. Recommendations and descriptions are based on first-hand observations of problem children in public school settings and emphasize individual programming. Discussions of specific problems in a variety of areas, suggestions for problem evaluation, and case history data are included. This book would be of interest to special educators, educators, and curriculum planners. (168 refs.) - J. K. Wyatt.

CONTENTS: Introduction: The Setting and Program; The Schools and Deviant Behavior; Special Education; What Is Special about Special Education? What Is Handicap? Education and Handicap; Teacher Preparation and Certification; The Selection of Teachers; Classroom Management and Behavioral Control; Permissiveness or Structure? The Classroom; Introduction to the Curriculum and Sample Lesson Plans; Language; Curriculum Materials; Spelling, Writing, Science, Social Studies, and Numbers; Evaluation for Educational Programming; Visual Processes; Auditory Processes; Non-Verbal Processes; Non-Verbal Processes: Writing; Associational Processes; Illustrative Studies of Individual Children.

- 2725 MACKIE, ROMAINE P., & *REYNOLDS, JULIA W. Compensatory education's contribution to the handicapped. *Education and Training of the Mentally Retarded*, 3(2):51-55, 1968.

Compensatory Education (Title I) and Provisions for Handicapped Children (Title VI) of

the Elementary and Secondary Education Act of 1965 provide a wealth of professional services for the MR and for those children functioning and performing at a low level of achievement due to cultural and environmental deprivation. If the substandard life situation of the deprived child could be alleviated or substantially improved, thousands of children would not require special education programs but could participate in a regular classroom setting. Title I has allowed the recruitment of educational specialists for the MR and the emotionally ill child. Nationally, the act has provided schools with guidance counselors, supporting personnel staff, laboratory teachers, and instructors for visually handicapped and hard of hearing children. Intensive teacher in-service training has been an integral phase of the program. Title I has broadened the spectrum of educational curriculum for handicapped children by providing more and improved enrichment programs, structured and organized summer activities, and tutoring and remedial assistance. Provisions are made under Title I for the allocation of funds to purchase special teacher's equipment, materials, and supplies and to engage in field trips which markedly contribute toward the handicapped child's attitude, motivation, and physical and mental well-being in school, at home, and in the community. (1 ref.) - S. Half.

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- 2726 ANDERSON, ROBERT M., HEMENWAY, ROBERT E., & ANDERSON, JANET W., eds. *Instructional Resources for Teachers of the Culturally Disadvantaged and Exceptional*. Springfield, Illinois, Charles C. Thomas, 1969, 304 p.

The emphasis of this comprehensive listing of instructional resources, which can be used for educational planning for culturally disadvantaged and/or exceptional children, is on remedial, adapted, experiential, and stimulation instructional materials and aids. Materials are classified by curriculum area rather than according to disability categories or etiological classification schemes because remediation should be based on the diagnosis of specific learning problems and because significant commonalities in the characteristics, needs, and problems of all exceptional children mean that many materials and methods are appropriate for more than one area of exceptionality. An introduction to each curriculum area includes a review of the ways in which it relates to exceptional and disadvantaged children, and an outline of its primary objectives, classroom implications,

broad areas of materials, and need. The format for each suggested instructional material includes a description and data on the publisher, author, cost, interest, and difficulty level. This book would be of interest to educators, teachers of the culturally disadvantaged and exceptional, college students, university instructors, school administrators, and library personnel. (24 refs.) - J. K. Wyatt.

CONTENTS: Introduction and Overview; Mathematics; Reading and English; Listening and Speaking; Writing and Typing; Spelling; Social Studies; Science; Health, Safety, and Sex Education; Occupational Education and Work Study; Driver Education; Physical Education and Motor Learning; Art; Music; Crafts; Home Arts; Industrial Arts; Curriculum.

- 2727 EDMONSON, BARBARA, LEACH, ETHEL M., & LELAND, HENRY. *Social Perceptual Training for Community Living. Pre-vocational Units for Retarded Youth.* Kansas City, Kansas, University of Kansas Medical Center, 1967, 306 p.

This manual consists of 50 lessons prepared for adolescent EMR children to enhance their social comprehension. The script format follows slides, sound tapes, and drawings developed for seatwork practice. The comprehensive program has been researched and tested for teaching the MR to read, understand, comprehend, and respond to signals and cues that they must learn in order to live and work meaningfully in the community. The structured lessons are in the form of sub-units and may be modified as to their length. A variety of presentational methods can be employed to sustain the children's interest. A teacher may develop a practical, interesting, and comprehensive course in social perception with little pre-preparation by appropriate utilization of this kit. Since much behavior is guided by signals that are nonverbal, it is essential that social cues be clear and interpretable by the MR, and appropriate behavior in response to the signals be clarified. Motivation is a prime consideration and can be effectively attained by various methods and techniques. (6-item bibliog.) S. Half.

- 2728 BOWERS, LOUIS. The status of physical education for the retarded. In: American Association for Health, Physical Education and Recreation, & National Recreation and Park Association. *Physical Education for Handicapped Children: Proceedings of a Study Conference on Research and Demonstration Needs.* Washington, D. C., 1969, p. 1-6.

Findings have indicated that MRs are 2 to 4 years behind normal children in physical performance; however, their patterns of development closely follow those of normal children. Physical conditioning programs result in increased motor proficiency, IQ scores, and muscular strength. The results of different projects have indicated a need for more adequate facilities, better CA grouping, better training for instructors, special seminars, workshops, and more funds. After determining the desired behavior for a particular child, an individualized comprehensive program should be designed which stresses improvement in physical fitness, motoric and social traits, perceptual development, movement experiences, basic skills, eye-hand coordination, and spatial awareness. (9 refs.) V. G. Votano.

- 2729 NUGENT, TIMOTHY J. Precepts and concepts on research and demonstration needs in physical education and recreation for the physically handicapped. In: American Association for Health, Physical Education and Recreation, & National Recreation and Park Association. *Physical Education and Recreation for Handicapped Children: Proceedings of a Study Conference on Research and Demonstration Needs.* Washington, D. C., 1969, p. 20-25.

Research studies must focus on the needs and the abilities of the disabled individual with the programs deviating as little as possible from normal patterns. Through participation in physical activity, the person with a disability realizes self-identification, overcomes self-consciousness, develops self-confidence, and has the opportunity for self-evaluation. There is a need for greater involvement of individuals with disabilities in programs, more involvement of professional people, more functional programs, research programs which utilize conditioning techniques, and a comprehensive review of current programs. Studies in motivation, attitude, values, psychological factors, heart function, energy expenditure, and evaluation must stress a normal environment and normal activities with a few modifications for the handicapped. (No refs.) - V. G. Votano.

- 2730 WHEELER, RUTH HOOK, & HOOLEY, AGNES M.
Physical Education for the Handicapped.
 Philadelphia, Pennsylvania, Lea & Febiger,
 1969, 352 p. \$9.75.

The physical education programs for atypical children and adults included in this book were derived from sound theory and practical experience. Data on the history of therapeutic exercise and physical education, rehabilitation and habilitation, the essentials of the nervous system, and the relation between motor learning achievement and body image concept are presented. Exercises for muscular fitness and re-education are illustrated and analyzed, and the correct body mechanics of daily tasks, common postural deviations, and teaching methods for activity programs are discussed. Fifteen handicapping conditions and appropriate physical education activities for them are considered in detail. Physical education activities which can be performed by MRs include dance, low organization games and team activities, aquatics, body mechanics, and movement exploration. Activity selection should depend on individual motor ability, motor potential, ability to understand directions, and desire to socialize. This book would be of interest to educators, special educators, physical therapists, and parents of handicapped children. (168-item bibliog.) - J. K. Wyatt.

CONTENTS: Historical Review and Developing Trends in Adapted Physical Education and Therapeutic Exercise; Rehabilitation and Habilitation: Physical Medicine, Allied Medical Services, and Physical Education; Essentials of the Nervous System; Motor Learning and Body Image; Mechanical and Muscular Efficiency; Daily Tasks and Body Mechanics; Common Deviations of Posture; Adjustment Problems; Implementation of the Physical Education Program for the Exceptional Child; Processing the Integrated Program of Adapted Physical Education; Future Directions in Adapted Physical Education.

- 2731 BOSTON UNIVERSITY, SCHOOL OF EDUCATION.
 A final report: The development and evaluation of three types of physical education programs for educable mentally retarded boys. Boston, Massachusetts, 1967, 261 p. Mimeographed.

Progress in physical fitness, motor ability, and social adjustment was compared for 100 institutionalized EMR boys (CA 10 to 15 yrs) enrolled in 3 different, 15-week physical education programs: play oriented (PO), skill oriented (SO), and free-play activity (FP).

Physical fitness was measured by critical incident technique of testing such areas as sit-ups, pull-ups, and standing broad jump. Motor ability was measured by the Latchaw Motor Achievement Test and included areas such as basketball wall volley and volleyball wall volley. Social adjustment was measured with the Cowell Social Adjustment Index which assesses both positive and negative behaviors. The skill oriented group made the greatest gains in motor ability and isolated elements of physical fitness; there were no differences between the free-play activity and play oriented groups. While the children tended to make gains in social adjustment, these were not systematically related to any of the programs. (No refs.) - J. M. Gardner.

- 2732 GILES, MARIAN TAYLOR. Classroom research leads to physical fitness for retarded youth. *Education and Training of the Mentally Retarded*, 3(2):67-74, 1968.

It was the purpose of this study to examine the effects of 3 specific motivations on the psychomotor functions of 20 children in a prevocational unit of special education in Amarillo, Texas. The effects were measured by a pretest and posttest battery given over a 5-month interval. The instruments used were the Wechsler Intelligence Scale for Children, National Youth Fitness Tests, and the Practical Dexterity Board Test. The results supported the hypothesis that a structured program of physical education combined with 4 specific techniques of psychoeducational therapy would improve the psychomotor function, physical condition, and social behavior of mildly retarded adolescents. (9 refs.) - Journal abstract.

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- 2733 HECHT, ARTHUR M. The preparation and placement of the educable retardate (Work-study program). In: Stark, Edward S., ed. *Special Education: A Curriculum Guide*. Springfield, Illinois, Charles Thomas, 1969, Chapter 5, p. 147-168.

A work-study program for MRs begins in junior high prework-study classes which emphasize a citizen education core program in which academic materials in the areas of travel, communication, and preparation for employment are designed to meet individual mental abilities. At age 16, special education teachers, administrators, and psychologists make decisions about the readiness of a student for

work by evaluating academic achievement, emotional stability, social maturity, and physical development. The curriculum for working senior students is a core program which integrates subject matter according to the interests, needs, abilities, CA, and development of each child. MRs should be assigned to regular homerooms and should participate in regular classes whenever possible. The work-study program should consist of 3 hours of instruction in the morning, one hour for lunch, and 3 hours of work. The work-study teacher analyzes the student for placement by making an initial evaluation, studying the job situation, setting-up a job portfolio, re-evaluating the student's records, redetermining the student's goals, and setting-up an interview with a prospective employer. The work-study teacher makes weekly visits to on-the-job training sites to provide additional programs to increase skill training and/or improve job adjustment. A unit on job preparation through occupational information, an outline of an occupational information program, suggestions about job placement, and a table of materials and sources related to work-study programs for the MR are included. (11-item bibliog.) - J. K. Wyatt.

2734 CHATTANOOGA PUBLIC SCHOOLS. A review of the educable mentally retarded work-study program in the Chattanooga Public Schools. Allen, Oscar. Chattanooga, Tennessee, 1967, 33 p. Mimeographed.

Of 509 EMR students (IQ range 45-79, CA 16 yrs and older) enrolled in a research and demonstration work-study project in 3 public high schools in Chattanooga (Tennessee) between 1964 and 1967, 70 graduated and 309 participated in vocational rehabilitation. Curriculum emphasis was on "planning for vocational competency" and included instruction in language skills, number skills and concepts, health and safety habits, social studies, and occupational information. Pupil-clients held 45 different types of prevocational jobs. During the project period, the number of EMR high school graduates more than doubled, the number of students placed in prevocational jobs increased, and student delinquency declined. Among the vocational rehabilitation services provided were medical, psychological, and work evaluations. (61-item bibliog.) - J. K. Wyatt.

2735 VLASOVA, T. A. Ob obuchinii umstvenno otstalykh detei v zarubezhnykh stranakh (The education of mentally retarded children in foreign countries). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 5-23.

The education of MR children is reviewed with special emphasis on a comparison of the Russian state-controlled system of educating the MR pupil in socialistic countries with the unstructured approach to special education which is prevalent in western countries. In the Soviet Union, education of MR Ss is carried out in schools for MRs who are selected not only by IQ, but also by their learning capacity. This capacity is judged by psychiatrists, psychologists, and educational experts. (No refs.) - R. K. Butler.

2736 GLAVE, H. Vospitanie i obuchenie slaboumykh detei v GDR (Educating and teaching feeble-minded children in East Germany). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 24-44.

The relatively modern East German remedial schools and their future direction are presented in a short history of the 8-year remedial schools in Germany. The majority of MR Ss finish remedial school at age 15 years and are then enrolled in special vocational schools. At present, there is a lack of research; therefore, the East Germans are using international experiences. The work of Soviet researchers is published in "Spetsial'naya Shkola" and is then translated into German. This literature is the mainstay of the East German organizations in the field of MR. Requirements for the formal training program of teachers for remedial schools are outlined, and a statistical comparison of remedial schools in 1949 and 1964 is given in the appendix. (17 refs.) - R. K. Butler.

2737 PETROVA, V. G. Obuchenie umstvenno otstalykh detei v Germanskoi Demokraticheskoi Respublike (po lichnym vpechatleniyam) (Teaching mentally retarded children in the German Democratic Republic [from personal impressions]). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otshtalykh Detei za Rubezhom* (Schools for Mentally Retarded Children in Foreign Countries). Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 45-50.

In East Germany, a MR class composition might include 7 mildly MR Ss, 3 moderately MR Ss, and 1 SMR S. Students are grouped by age and by the severity of MR. A child is observed by a teacher and, after 6-8 days, is mixed in with a special group consisting of 10 Ss. One teacher instructs while 2 other teachers observe the children and take notes. Medical and psychological testing is also done. The pupils will leave the schools at 16 to 18 years of age; the mildly MR will have completed 10 grades, while the moderately MR will have completed 8 grades and the SMRs 6 grades. After third grade, boys and girls work together learning a trade. From the eighth to tenth grades, Ss with mild MR work one 4-hr day/week in an industrial enterprise; studies are now being done to determine probable success or failure with moderately and severely MR Ss. (No refs.) - R. K. Butler.

2738 FID'OV, D. Sistema obucheniya, vospitaniya i otbora umstvenno otstalykh detei v Bolgarii (System of educating, teaching, and selecting mentally retarded children in Bulgaria). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otshtalykh Detei za Rubezhom* (Schools for Mentally Retarded Children in Foreign Countries). Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 51-57.

Developments in the education of MR Ss in Bulgaria from 1907 to the mid-1960's is presented. MR children (CA range 4 to 7 yrs) attend special kindergartens where they are separated into groups by the level of their mental development and by their age. This classification is maintained throughout the school life which continues for 8 years after kindergarten. Selection is made before May 30 and school classes start September 15 for all grades; they finish by May 31 for first through fourth and eighth grades and by June 15 for all others. Seventh and eighth graders work 4 and 5 hours/week, respectively, in industrial enterprises. A uniform teaching plan covers the subjects taught and the time

spent on each for all 8 yrs. During these 8 years, MR Ss spend an average of 30 hours/week in formal education. (No refs.)

R. K. Butler.

2739 MASYUNIN, A. M. Dom dlya gluboko umstvenno otstalykh mal'chikov v Sofii (po materialam Todora Popova) (Home for severely mentally retarded boys in Sofia [According to Todor Popov's material]). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otshtalykh Detei za Rubezhom* (Schools for Mentally Retarded Children in Foreign Countries). Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 58-61.

The Sofia Home for SMR boys has age limits from 10- to 20-years old; those from 10 to 16 years work 2 to 4 hours/day, while those 16 to 20 years work 5 to 7 hours/day. Formal education of SMR Ss has not given positive results; therefore, education is not done during the work periods. The trainees are generally taught about their daily experiences. Thus, the pupils learn to communicate with each other. SMR boys are trainable for simple tasks and should be used in the work force. (No refs.) - R. K. Butler.

2740 BEDOR, E. Voprosy izucheniya, obucheniya i vospitaniya umstvenno otstalykh detei v Vengrii (Problems of studying, teaching and educating mentally retarded children in Hungary). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otshtalykh Detei za Rubezhom* (Schools for Mentally Retarded Children in Foreign Countries). Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 62-82.

New advances in the development of Hungarian defectology during the past 14 years include a 9-year education program with separate schools for TMRs and SMRs, curriculum for the program, and a new type of school for remedial school graduates--a remedial vocational (industrial and agricultural) school for further training. (No refs.) - R. K. Butler.

2741 ZAMSKII, KH. S. Vospitanie i obuchenie umstvenno otstalykh detei v Vengrii (polichnym vpechatleniyam) (Education and teaching of mentally retarded children in Hungary [from personal impressions]). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 83-87.

Hungarian remedial schools have a system of preparatory classes (experimental, transitional, and preparatory) preceding the first class. In these classes, MR diagnosis is confirmed, the child's possibilities are studied, the subsequent system of education for each child is noted, and work habits are cultivated. The latest advance in Hungarian remedial schools is the strong emphasis being placed on vocational education. Two schools (one with 410 Ss; the other with 636 Ss) are discussed, as well as the 4-year teacher training institute. (No refs.) - R. K. Butler.

2742 POPESCU, G., & RADU, G. Sistema spetsial'nykh uchrezhdenii dlya umstvenno otstalykh detei v sotsialisticheskoi respublike Rumynii (System of special institutes for mentally retarded children in the socialist republic of Rumania). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 88-96.

Rumania has set up a unique system of schools for MR children in which kindergartens for 4- to 7-year olds remain open 9 hours/day and in which there are 7-year remedial schools for 7- to 16-year olds. After the remedial schools, there are 3 special vocational boarding schools which include one for MRs who finished remedial school, one for MRs who did not finish remedial school by the age of 16 or who did not pass 2 to 3 classes in a regular school, and one for those Ss who had not progressed at all by the time they became 14 years old. (No refs.) - R. K. Butler.

2743 MESHCHERYAKOV, A. I. Ob obuchenii umstvenno otstalykh detei v sotsialisticheskoi respublike Rumynii (Educating mentally retarded children in the socialist republic of Rumania). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 97-100.

The educational system for MR children in Rumania and research in the fields of MR and related subjects at the University of Cluj are discussed. The works of Russian defectologists are systematically translated and published in Rumania since Russian practical experience and achievements are widely used. (No refs.) - R. K. Butler.

2744 DUL'NEV, G. M. Vspomogatel'nye shkoly v Pol'skoi Narodnoi Respublike (Remedial Schools in the Polish People's Republic). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 101-109.

The 2 types of remedial schools in Poland are the basic 7-year remedial school and the 2- to 3-year vocational school. If a child has an IQ of less than 75-77 on the Stanford-Binet scale, he is usually placed in a remedial school. (His personality and psychological traits are also considered.) The "method of centers" is used in educating the MR; subjects, such as "my family," "our street," and "nature around us," are used because the MR is familiar with these social and natural surroundings. In 1960, 2% of all school-age children in Poland were considered MR. (No refs.) - R. K. Butler.

2745 DOROSHEVSKAYA, YA. Podgotovka kadrov dlya vspomogatel'nykh shkol v institute spetsial'noi pedagogiki v Varshave (Preparation of remedial school faculties in the Institute for Special Education in Warsaw). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 110-114.

A 2-year education plan for teachers who will work in remedial schools in Poland is available for regular school teachers. To qualify,

an elementary school teacher with 2 years of practical teaching experience must pass an entrance examination. The first semester includes familiarization with organizations for mother and child preservation and special schools for exceptional children. During the second semester, a teacher decides which area he prefers, and once a week, he attends a lecture with a specialist in his chosen field. In the second year of study, for one week/month, the teacher visits and teaches in a special institute in his field. After passing final examinations, the teacher must write and defend a thesis. (No refs.)

R. K. Butler.

2746 GANNO, VIL'YAM, & PREDMENSKII, VLADIMIR. Razvitie spetsial'nykh shkol dlya umstvenno otstalykh detei v Chekhoslovatskoi sotsialisticheskoi respublike (Development of special schools for mentally retarded children in the Czechoslovakian Socialist Republic). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 115-125.

In Czechoslovakia, there are special schools for MR children with defects in hearing, speech, vision, and physical handicaps. Textbooks, vocational schools, and teacher education have progressed steadily since 1871; however, new journals and books which will help the teachers of MR Ss are needed. Historically, MR has increased in Czechoslovakia; in the period 1936-1937, there were 116 schools with 8,672 pupils, while in 1964-1965, there were 1,028 schools with 57,863 pupils and 28 vocational schools with 3,138 pupils. (No refs.) - R. K. Butler.

2747 KUZ'MITSKAYA, M. I., LUR'E, N. B., & PETROVA, V. G. Vospitanie i obuchenie umstvenno otstalykh detei v Chekhoslovakii (Education and teaching mentally retarded children in Czechoslovakia). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 126-136.

Remedial schools in Czechoslovakia usually have 3 departments to serve EMRs, SMRs, and MR children with poor vision. A 3-year vocational school education includes pay for students; first-year students earn 70 koruna (\$9.72)/month, second-year students earn 100

koruna (\$13.89)/month, and third-year students earn 80% of an adult's salary. Weekly teaching plans are given. Czechoslovakian literature on MR is greatly enlarged by translations from Russian, Polish, German, and Hungarian MR journals. (No refs.)

R. K. Butler.

2748 NOVAKOVIC, B. Obuchenie i vospitanie umstvenno otstalykh detei v Yugoslavii (Teaching and educating mentally retarded children in Yugoslavia). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 137-148.

The history, educational system, placement of MRs into remedial schools, teaching plans, and vocational and teacher education in Yugoslavia are discussed. Selection into remedial schools is through verbal and non-verbal tests including the Binet, Wechsler, and Raven. In 1937-1938, there were 5 remedial schools and 95 classes for MR children in regular schools; in 1963, there were 23 remedial schools with 2,365 pupils and 217 classes in regular schools with 2,795 pupils. In addition, there were 4 institutes for productive education of young people which had 253 students, 15 children's homes with 1,982 children, 11 diagnostic centers, and several medical institutes for MR children. (No refs.)

R. K. Butler.

2749 ZAMSKII, KH. S. Nekotorye voprosy spetsial'nogo obucheniya umstvenno otstalykh detei v kapitalisticheskikh stranakh (po materialam YuNESKO) (Some problems of the special education of mentally retarded children in capitalist countries [According to UNESCO data]). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 149-157.

Problems involved in educating MR children in the non-Soviet world are reviewed. Those countries in which education for MR Ss is not compulsory are listed. Placement of MR children in remedial schools, the type of schooling, and the education of teachers are discussed. The 23 recommendations worked out by the XXIII International Conference on National Education are listed. (No refs.)

R. K. Butler.

2750 LURIYA, A. R. Nekotorye voprosy organizatsii obuchenni umstvenno otstalykh detei v Norvegii (Some problems of the organization of the education of mentally retarded children in Norway). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 158-164.

The placement of MR children in remedial schools in Norway is compared with that of England; in some English counties, at 11 years of age, a child is given a series of psychometric tests which determine the course of the rest of his life. In Norway, teachers, psychiatrists, psychologists, and special social workers are involved in placement, and there is a central registry for all students which aids in the planning for the future. Norwegian institutions include those for EMRs, those for SMRs, and those for PMRs who are incapable of learning. In addition, there are children's day care centers which are open from 7 AM to 7 PM. (No refs.)

R. K. Butler.

2751 LUBOVSKII, V. I. Sistema obucheniya vospitaniya i trudovoi podgotovki umstvenno otstalykh detei v Danii, Shvetsii i Gollandii (System of teaching, education, and vocational preparation of mentally retarded children in Denmark, Sweden and Holland). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 165-176.

A state-of-the-art review from reports made by the president of the United States Commission on Problems of MR of studies of Denmark, Sweden, and Holland in 1962 is given. Denmark and Sweden do not use EMR or TMR terminology but instead debiles (IQ of 50-75), imbecile (IQ of 35-50), and idiot (IQ of 35 or less) as does the Soviet Union. In the Netherlands, debiles have an IQ of 50-80, imbeciles an IQ of 20-60. In Denmark, 1% of the population and, in Sweden, 1.8% of the population are MR; however, Ss with schizophrenia are included in the figures. There is compulsory education of MRs in Denmark from 7 to 21 years and in Norway from 7 to 23 years. The borderline or slow learners have special classes. Teachers in Denmark study 4 months; in Sweden, 5 months.

In 1946, 147 schools served 16,510 Ss; in 1951, 206 schools served 25,888 Ss; and in 1962, 330 schools served 40,000 Ss in the Netherlands. Compulsory education for EMRs lasts from 7 to 15 years; for imbeciles, it lasts from 6 to 18 years of age. In all 3 countries, much attention is being paid to education, teacher education, and vocational education of MR children. (No refs.)

R. K. Butler.

2752 LUBOVSKII, V. I. Obuchenie umstvenno otstalykh detei v Anglii (Teaching mentally retarded children in England). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 177-187.

The placement of MR children in schools and the structure, problems, and programs of schools for MR children in England are discussed. Schools for EMR children in England correspond to USSR remedial schools and enroll EMRs with IQ levels of 50 to 70. In 1962, there were 32,000 Ss in 340 schools (this does not include those Ss in special classes at normal schools). The mean average percentage of MR children in the total children's population is 0.8%. One MR school in Birmingham and a training center in Manchester are described. (4 refs.) - R. K. Butler.

2753 ZAMSKII, KH. S. Vospitanie i obuchenie umstvenno otstalykh detei v Anglii (Educating and teaching mentally retarded children in England). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 188-192.

A brief review of MR education in England from 1945 to 1959 is given. For 1945-1955, the Ministry of Education estimated the total number of MR (educationally neglected and underdeveloped) to be 11% of the total 5- to 15-year-old group. As of 1955, it was estimated that 50% of those MRs needing special education did not receive it, while in 1959,

this had decreased to about 30%. (2 refs.)
R. K. Butler.

2754 ZAMSKII, KH. S. Vospitanie i obuchenie umstvenno otstalykh detei vo Frantsii (Educating and teaching mentally retarded children in France). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 193-197.

In 1960, of 225,000 French MR children (4.5% of all school-age children), less than 25% (50,000) were receiving the needed special education. In France, an MR is one who, if under 9, is 2 years behind his peers and, if over 9, is 3 years behind in school knowledge and development. Binet-Simon tests, with others, are used as diagnostic tools. French special schools for MR include classes in normal schools, daily remedial schools,

boarding remedial schools, national vocational training centers, and special classes and boarding schools in hospitals. (2 refs.)
R. K. Butler.

2755 PETROVA, V. G. Obzor literaturi po obucheniyu umstvenno otstalykh detei v SSHA (Literature review on the education of mentally retarded children in the USA). In: Vlasova, T. A., & Shif, Zh. I., eds. *Shkoly dlya Umstvenno Otstalykh Detei za Rubezhom (Schools for Mentally Retarded Children in Foreign Countries)*. Moscow, Union of Soviet Socialist Republics, Prosveshchenie Press, 1966, p. 198-211.

A literature review on the education of United States MR children up to 1964 is given. The randomness of the quality of education and the reasons for it are discussed. The American Association on Mental Deficiency is one of the main literature sources which set forth the basic advances of work with the MR. (10 refs.) - R. K. Butler.

TREATMENT AND TRAINING ASPECTS - PSYCHO-SOCIAL

2756 GRAY, ROBERT M., & KASTELER, JOSEPHINE M. The effects of social reinforcement and training on institutionalized mentally retarded children. *American Journal of Mental Deficiency*, 74(1):50-56, 1969.

This paper reports an evaluation of the effects of a program in which older men and women were employed as foster grandparents to give love, individual attention, and training to a group of institutionalized MR children. Treatment consisted of individualized training, using methods of imitative learning and reinforcement. Focus in this paper is on the development of social competence. Findings led to the conclusion that institutionalized retarded children can reach higher levels of social competence if

given individual attention and special training and that teachers need not be professionally trained, but can be volunteers or lower paid workers. (29 refs.) - *Journal abstract*.

University of Utah
Salt Lake City, Utah

2757 GARDNER, WILLIAM I. Use of punishment procedures with the severely retarded: A review. *American Journal of Mental Deficiency*, 74(1):86-103, 1969.

Behavior treatment procedures involving aversive consequences have been used with increased frequency in work with the severely

and profoundly retarded. Review of these punishment studies suggests a cautious conclusion that such procedures may produce desirable behavior change. In addition, there is some evidence that side effects of negative emotional states and disruption of social relationships are not necessary results of punishment techniques. (60 refs.)

Journal abstract.

University of Wisconsin
Madison, Wisconsin 53706

2758 THOMSON, ROBERT G. The use of operant conditioning techniques in the habilitation of disturbed adolescent retardates. In: Fuzessery, Zoltan, ed. *New Frontiers in Psychiatric Technology*. Sacramento, California, National Association of Psychiatric Technology, 1969, p. 36-46.

Of 71 disturbed adolescent institutionalized MR boys (IQ range 40 plus, CA range 13 to 24) who participated in an operant conditioning program, 22 were referred for community placement, 7 were promoted to the hospital's co-ed ward, the balance are awaiting administrative action and only 3 did not improve. All Ss were relatively free of any physical or organic dysfunction and had some type of noticeable disturbed behavior. The 16-hour-a-day program was designed to shape desirable behavior through the use of rewards and a token economy system. Everyday experiences were used as teaching and learning experiences. Learning experiences began at the ward level and progressed, as the child improved, to the school, skilled training assignments, and community activities. Negative reinforcement was used in the form of fines, deprivation, and restriction to the ward. The boys progress through 3 levels (junior, intermediate, and senior levels) with privileges on each level being greater than those on the preceding one and the token cost of goods decreasing as the level increased. To remain at the intermediate and senior levels, a S must maintain set standards of behavior. (No refs.) - J. K. Wyatt.

2759 KILBURN, KENT. Implications of using operant conditioning. In: Fuzessery, Zoltan, ed. *New Frontiers in Psychiatric Technology*. Sacramento, California, National Association of Psychiatric Technology, 1969, p. 47-55.

Operant conditioning for the mentally ill and MR aims to develop these individuals to their fullest potential and places primary

emphasis on the development of acceptable behavior and secondary emphasis on the reduction of inappropriate behavior. Operant conditioning is based on the idea that consistent and systematic reinforcement of acceptable behavior will strengthen it and slowly eliminate inappropriate behaviors which are no longer profitable. In the operant conditioning program at Porterville State Hospital (California), psychiatric technicians are trained to monitor patients' behavior and effect behavior change. The program emphasizes the use of appropriate rewards and the principles of immediacy, consistency, contingency, and schedules of reinforcement. Negative reinforcement is used when necessary and is applied in a 2-stage process in which inappropriate behavior is pointed out and an appropriate substitute suggested. The program emphasizes self-care skills and elimination of behavior problems. Since the use of naturally occurring rewards has several serious limitations, a token system of rewards was adopted. Operant conditioning is not a fad, but rather a body of knowledge which can be applied to a wide variety of hospital and community populations. (No refs.) - J. K. Wyatt.

2760 DODGE, MADELINE R., & HARRIS, FLORENCE R. Use of reinforcement principles with autistic children. In: Washington. Institutions Department. *Proceedings of the Ninth Annual Research Meeting*. Olympia, Washington, *Research Report*, 2(2):82-84, 1969.

The results of 2 studies indicate that systematic adult social reinforcement and primary reinforcement in a well-planned and rich stimulus environment can help to modify the behavior of 2 autistic children. During a 7-day observation period, the operant level of appropriate and inappropriate behaviors was determined for a 7 1/2 year-old boy, and adult attention elicited by these behaviors was recorded. Social reinforcement was then instituted by the staff; appropriate behaviors were given immediate attention and inappropriate behaviors were ignored. In addition, the S received individual training in academic work and active play for one-hour-a-day, 3 days a week. Social and primary reinforcements were used during the training sessions. Baseline data indicated that appropriate behavior occurred 27% of the time, inappropriate behavior was noted 49% of the time, and adults gave their attention to either type of behavior 50% of their time. During the 21-day reinforcement period, appropriate behavior was recorded an average of 72% of the time and inappropriate behavior occurred 2% of the time. Appropriate verbal behavior rose from a baseline of 1% to 45%,

and from one-word intervals to as many as 6 or 8 word intervals. When similar techniques were used with a mute autistic boy (CA 4 yrs), the amount of time (55%) he spent playing with a string was virtually eliminated. (3 refs.) - J. K. Wyatt.

- 2761 PETERSON, ROBERT F. Some experiments on the organization of a class of imitative behaviors. *Journal of Applied Behavior Analysis*, 1(3):225-235, 1968.

Diverse motor and verbal imitative responses were taught to a profoundly retarded 12-year-old girl. At mealtime, food was delivered contingent upon correct responses occurring within 30 seconds of the demonstration. In experiment 1, non-reinforced responses could be extinguished under massed stimulus presentation; however, they were performed when interspersed among reinforced responses. In experiment 2, the S failed to perform all non-reinforced imitative or non-imitative responses regardless of the similarity to the reinforced stimuli. Experiment 3 demonstrated that non-imitative and imitative responses are performed under conditions of massed and interspersed stimulus presentation. In experiment 4, non-imitative responses declined as soon as reinforcement was discontinued for the imitative responses. These studies demonstrated that imitative responses can serve as a functional response class and may be part of an even larger response class

which includes non-imitative responses as well. (12 refs.) - J. M. Gardner.

University of Illinois
Champaign, Illinois 61820

- 2762 ZIGLER, EDWARD. Outer-directedness in the problem solving of retardates. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 15, p. 123-127.

The outer-directness hypothesis suggests that the distractibility often attributed to MR children is actually a style of problem solving which emanates from their experiential histories. The high incidence of failure experienced by MRs causes them to distrust their own solutions and to look for action guides in their immediate environment. Outer-directness in MRs, therefore, is characterized by an oversensitivity to external models and a lack of spontaneity and creativity. Its development appears to depend on the cognitive level of the individual and on the degree of success experienced by using available cognitive resources--the lower the mental age, the greater the degree of outer-directness. Outer-directness is learned early and appears to generalize to many external stimuli. This generalization requires the child to attend to a wide variety of stimuli since he seeks problem solving clues from the environment rather than from his own experience. (6 refs.) - J. K. Wyatt.

TREATMENT AND TRAINING ASPECTS - OCCUPATIONAL

- 2763 HUNT, JAMES G., & ZIMMERMAN, JOSEPH. Stimulating productivity in a simulated sheltered workshop setting. *American Journal of Mental Deficiency*, 74(1):43-49, 1969.

Productivity in "exit ward" patients, participating in a simulated workshop setting, was examined as a function of introducing a bonus pay procedure. Work units completed/hour served as the dependent variable, and coupons redeemable for canteen items served as reinforcers. The bonus procedure significantly increased group productivity above that previously obtained under non-bonus conditions

and differentially maintained productivity at values consistently higher than those obtained during temporally adjacent non-bonus periods. While these results could have been accounted for exclusively on the basis of the bonus procedure, they could also have been influenced by verbal instructions given in conjunction with that procedure. (9 refs.) *Journal abstract.*

Ball State University
Muncie, Indiana 47306

2764 LEVINE, MILTON. A sheltered workshop experience for the mentally retarded. In: Stark, Edward S., ed. *Special Education: A Curriculum Guide*. Springfield, Illinois, Charles Thomas, 1969, Chapter 6, p. 169-187.

Sheltered workshop training programs conducted at the Association for the Help of Retarded Children Vocational Training and Educational Centers (New York) include bench assembly, machine operations, a greenhouse program, shipping and receiving, merchandise handling, office practices, tool shop, custodial and janitorial work, cafeteria, and child care. Job training is specific and aimed at producing flexible work concepts which can be easily translated from one occupational area to another. The workshops provide a relatively anxiety-free environment which is conducive to systematic, vocationally-structured learning and to trial-and-error problem solving. Descriptions of each training program and of the equipment and training titles used in it are included. Functional work categories which conform to those in the *Dictionary of Occupational Titles* are listed, and descriptions of the work performed under each category, its requirements, and training possibilities are included. (No refs.) - J. K. Wyatt.

2765 JEFFERSON COUNTY, KENTUCKY COUNCIL FOR RETARDED CHILDREN. *The Sheltered Workshop and Occupational Training Center: Final Report, Grant Number 1547-D*. Whitmer, Jack N. Louisville, Kentucky, 1967, 69 p.

The effectiveness and comprehensiveness of a vocational rehabilitation service was demonstrated in an occupational training center. The Ss were between the ages of 16-44 years and had IQs ranging from 50-70. Evaluation took psychological and vocational factors and work samples into consideration. The staff included a director, social workers, production superintendent, rehabilitation counselor, and special instructors. Work contracts were obtained by a workshop committee, mailing brochures, telephoning, and retired citizens. Production problems included shortage of contracts, proper loading docks, and movement of materials. (No refs.) - V. G. Votano.

2766 BADELL-RIBERA, ANGELES, SIEGEL, MEYER S., & SWINYARD, CHESTER A. Vocational rehabilitation case load and potential of patients with spina bifida and myelomeningocele. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 134-147.

A re-evaluation of 27 patients with spina bifida and myelomeningocele was based on a review of initial admission data which emphasized medical and functional status and on follow-up data obtained from 3 to 15 years after initial admission. Changes in the condition of the Ss were mainly due to the natural development of their disability since none had received surgical treatment for hydrocephalus and orthopedic surgery had been minimal. Renal function had deteriorated in 7.4% of the Ss. The severe orthopedic deformities in Ss with the greatest neurological defects resulted in greater physical incapacitation over the years, and the number of patients who had poor orthopedic status was about 10 times greater at time of re-assessment. Thirty-seven percent evidenced emotional disturbance, 22.2% were obese, and 51.8% had decubitus ulcers. Of the 6 Ss who had intellectual deficits, 5 had IQs below 80 and poor social and educational adjustment. Primary grade education had been provided by a home teacher (10 Ss), in special classes (2 Ss), and in regular school (14 Ss). Educational and vocational progress was closely correlated with intellectual ability. (No refs.) - J. K. Wyatt.

2767 TIZARD, JACK. Rehabilitation and employment of the severely retarded: An overview. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 27, p. 209-215.

To plan comprehensive services for SMRs (IQ below 50) which meet their rehabilitation and employment needs, data on prevalence, associated handicaps, family problems, institutional care, and currently available programs are needed. The prevalence of SMR in 10- to 19-year-old children in England is 36/1,000; true prevalence rates for older

and younger age groups are not available. Although data on trends in prevalence are lacking, the incidence of handicapping conditions (at birth) which contribute to SMR appears to be declining while expected life span is increasing. Two-thirds of the SMRs who reside at home and 1/3 of those who are institutionalized do not have major disabilities apart from MR. Sixty-six percent of the families with an MR child at home and 45% of the families with an institutionalized MR child had 3 or more family problems, and many received little support from social agencies. Research on the education of the SMR indicates that their potential for education and employment is greater than had been previously thought and that suitable education enhances intellectual growth, communication, and emotional maturity. Most SMRs can be usefully and happily employed in sheltered workshops. Research needs in this area include intensive social and psychological studies of individuals and small groups, large-scale epidemiological surveys of prevalence of handicapping conditions, and longitudinal studies of development. (18 refs.) - J. K. Wyatt.

- 2768 BADELL-RIBERA, ANGELES, SWINYARD, CHESTER A., GREENSPAN, LEON, & DEEVER, GEORGE C. Spina bifida with myelomeningocele evaluation of rehabilitation potential. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 88-99.

Of 86 patients (CA range 7 mos to 26 yrs) with spina bifida and myelomeningocele, 19 had no lower extremities motor power but good motor power in the trunk and above (Group I); 27 had hip flexion and adduction motor power and some knee extension (Group II); 7 had motor power for hip flexion and adduction, knee extension, and ankle flexion and dorsi-flexion (Group III); 21 had good hip motor power except for extension and adduction as well as good knee extension and flexion, and some plantar flexion, ankle eversion, and toe movement (Group IV); and 12 had functionally normal motor power in the lower extremities with some loss in the perineal area (Group V). Analysis of acquired skeletal or soft tissue deformities revealed no cases in Group V and the highest incidence among Ss in Group II. Group II Ss had frequent cases of hip dislocation and paralytic and malposition deformities. All Ss were incontinent, and Ss in Groups IV and V frequently had skin lesions which interfered with their total function. Intellectual function was within the average

or educable MR range. In Ss with known arrested hydrocephalus, there was usually a large discrepancy between verbal and performance scores on the Wechsler Intelligence Scale for Children. To provide a suitable social environment, continuous parental education programs should be provided. These programs should be designed to help parents achieve practical management and treatment goals and to lighten the family burden. Spina bifida patients require a rehabilitation program which prevents progressive disability, prepares them for admission to school facilities as children, and prepares them for vocational or professional training as adults. (20 refs.) - J. K. Wyatt.

- 2769 MORDOCK, JOHN B., & FELDMAN, RUTH C. A cognitive process approach to evaluating vocational potential in the retarded and emotionally disturbed. *Rehabilitation Counseling Bulletin*, 12(3):136-143, 1969.

Despite the fact that cognitive functioning is an important area of concern, there are no valid and reliable instruments for its measurement. Traditional intelligence and personality tests have been unable to point to the cognitive factors which make for adequate vocational adjustment. In recent years, perceptual processes were studied in an attempt to elucidate cognition and highly stable patterns within different individuals for discovering and processing information were discovered. This may prove to be a useful approach to the study of cognition. (24 refs.) J. M. Gardner.

- 2770 SOUTH CAROLINA. RICHLAND COUNTY SCHOOL DISTRICT ONE. Coordinated Program of Special Education and Vocational Rehabilitation Services for the Mentally Retarded: A Study and Demonstration of the Effects of a Special Education-Vocational Rehabilitation Program for Secondary School Retarded Adolescents. Final Report of Project RD-1649. Corder, W. Owens. Columbia, South Carolina, 1967, 104p. Mimeographed.

The major purposes of a special education research and demonstration project were to identify and serve secondary school MR adolescents who demonstrated potential for successful vocational rehabilitation, to accelerate rehabilitation services for the severely handicapped, and to enhance pupil-client interest in self improvement. The 127 Ss (mean IQ 68.5; CA range 13-21 yrs; mean social age 12.8 yrs) were given an extensive psychological evaluation, and approximately

2/3 were provided with comprehensive pre-vocational services. A team approach to evaluation, counseling, job training, and job placement was used. Over the 3-year period, 19 Ss were successfully placed in jobs, and 26 dropped out of school. The goals of the curriculum were the development of social competence, the improvement of the self-concept, and the enhancement of social adjustment and motivation. The curriculum included special work in arithmetic, language arts, health, science, personal and social adjustment, social studies, and physical education. Ss spent part of each day at an evaluation center where their eye-hand coordination, work tolerance, ability to follow directions, concentration, and overall job potential were evaluated. The major aim of an educational-vocational rehabilitation project should be to prepare each client for maximum job performance and help him become a contributing member of his community. Since the length of time required to accomplish this aim varies with the individual, separation from the project should not occur until the staff determines that a client is ready for placement. (20-item bibliog.) - J. K. Wyatt.

2771 KENTUCKY. REHABILITATION SERVICES BUREAU AND EDUCATION DEPARTMENT. Co-ordinated Program of Vocational Rehabilitation and Special Education for the Mentally Retarded in Harlan County, Kentucky. Final Report of Project 1522-D-67-C2. Harlan, Kentucky, 1967, 46 p. Mimeographed.

One hundred and nine high school students and dropouts were successfully rehabilitated during the first 3 years of a cooperative demonstration project between the Bureau of Rehabilitation Services and the school systems in Harlan (Kentucky). The project provided vocational counseling and job training for referred students (CA range 15-21 yrs; IQ range 40-85) in regular and special education classes and for former students. Among the project services were psychological, social, and medical evaluations; vocational counseling, planning, and evaluation; job development training and pre-employment work experience; special education programs designed to maximize educational and social development and vocational readiness; prevocational training; work-study programs; and formal vocational training. The program operated a Work Evaluation Center which provided an atmosphere similar to a real work setting with real work incentives. Rehabilitation procedures included initial referral, a preliminary case study, psychological and medical examinations, the development of an outline of appropriate special education and rehabilitation activities, case conferences with the

project staff, counseling with clients' parents, vocational training, job placement, and follow-up. Clients were selected for the program because it seemed unlikely that they could attain satisfactory work adjustment without it and because they demonstrated that their potential for learning new vocational skills and for personal and social development was greater than that predicted by test scores. (7-item bibliog.) - J. K. Wyatt.

2772 KENTUCKY. REHABILITATION SERVICES BUREAU AND EDUCATION DEPARTMENT. An organized Coordinated Program of Vocational Rehabilitation and Special Education for the Mentally Retarded. Final Report of Project RD-1524-D. Prestonburg, Kentucky, 1967, 40 p. Mimeographed.

The purpose of a 3-year coordinated program between the Kentucky Bureau of Rehabilitation Services and the Floyd County Schools was to provide joint education and rehabilitation services for MR students and school dropouts. The project goal was to provide experiences, training, education, and any other services needed to help clients develop into independent, contributing citizens capable of full-time employment. The project included adolescents from 15 to 25 years of age with IQs within the 50-85 range. Of 190 Ss who were placed in employment after participating in the program, 74 received on-the-job training, 34 were trained at vocational schools, and 86 received counseling, guidance, and placement only. Of the 1,036 referrals to the program, 526 were administered psychological tests. Of 261 Ss accepted for services, 20 were not rehabilitated, and 51 cases were not completed at the close of the project. IQ alone was not a predictor of rehabilitation success. Rehabilitation results were also influenced by: family, home, and economic background; academic performance; physical or emotional problems; motivation; and social behavior. (6 refs.) - J. K. Wyatt.

2773 KENTUCKY. REHABILITATION SERVICES BUREAU AND EDUCATION DEPARTMENT. Co-ordinated Program of Vocational Rehabilitation and Special Education for the Mentally Retarded in Rockcastle County, Kentucky. Final Report of Project RD-1523. Mt. Vernon, Kentucky, 1967, 44 p. Mimeographed.

A cooperative demonstration project between the Bureau of Rehabilitation Services and the Rockcastle County School System (Kentucky) provided vocational counseling and job training for MRs between the ages of 15 and 21. Of 408 clients who applied for services during

the 3-year project (mean IQ 69.93; mean educational level 9.1 yrs; 258 males, 123 females), 116 were successfully rehabilitated, 131 were not formally accepted for services, 151 cases are still open, and 10 clients were not rehabilitated. Students were referred to the project from special education classes or by counselors or teachers. Specific project services included: medical, psychological, and social evaluations; vocational counseling, planning, and evaluation; special education classes designed to enhance development, vocational readiness, and training in specific vocational areas; work-study programs; and formal vocational training. Most on-the-job training placements were on farms; however, a significant number of Ss were trained as machinists. Successfully rehabilitated Ss were employed in a wide variety of occupations. Obstacles to the project were the lack of training facilities and jobs. When the counselor has the support of the business community, this type of program can help in the vocational rehabilitation of a significant number of MRs, even in an area of chronic high unemployment. (3-item bibliog.)

J. K. Wyatt.

2774 NORTHERN KENTUCKY GOODWILL INDUSTRIES REHABILITATION CENTER. Northern Kentucky Goodwill Industries Occupational Training Center for the Mentally Retarded. Final Report of RD-1695-SD. Covington, Kentucky, 1968, 84 p. Mimeographed.

One hundred (mean IQ 63; mean CA 19.4 yrs) of 176 clients who applied to the Occupational Training Center for the MR of Northern Kentucky were given assistance which included work evaluation. Of these 100, 43 obtained employment in the open labor market, 12 are employed in sheltered workshops, and 24 are in vocational preparation programs. None of the 76 clients (mean IQ 78; mean CA 18.8 yrs) who did not receive services are employed in sheltered workshops, and 16 are employed in competitive industry. The project provided work evaluation, personal and work adjustment training, on-the-job training, guidance, and selective job placement. Among evaluated clients, males were significantly ($p < .001$) easier to place than females. Prior to participation in the program, the typical applicant had about a 20% chance of obtaining competitive employment. After receiving the services of the Center, chances for employment in competitive industry rose to 43%, and chances for some type of employment were at least 55%. Most applicants had completed 7 years of school and tended to be near the upper limits of intelligence for MRs. (No refs.) - J. K. Wyatt.

2775 MEMPHIS DEPARTMENT OF INSTRUCTION. Co-ordinated program of vocational rehabilitation and special education services for the mentally retarded: Final Report of Project No. RD-1682. Memphis, Tennessee, 1968, 34 p. Mimeographed.

A city education department 4-phase demonstration project to evaluate, train, employ, and follow-up 16- to 21-year old EMRs was successful in its purpose to set realistic vocational goals and make a smooth transition from school to work. Staff consisted of a project director and associate, vocational counselor, special education teachers, psychometrist, and a vocational rehabilitation staff member. Evaluation and Training Centers were set-up in vacant areas of 2 grade schools, and EMRs were referred by schools and agencies and evaluated during the school year by the Vocational Rehabilitation Agency. If accepted, the EMR entered phase 1 in an Evaluation and Training Center and his vocational talents were observed while training in services (food, sewing, clerical, shoe repair, hotel, custodial, stock clerk, wood working, and crafts). After evaluation, the EMR entered phase 2, which was designed to promote good work and emotional habits and was supervised by vocational counselors. Phase 3 consisted of training in a specific occupation, and phase 4 entailed full-time employment in the community with a counselor being vital to transition success. In 3 years, 115 of 152 EMRs were permanently employed (76%), and businesses are more open to the idea of employing EMRs. A similar program has now been adopted in all 28 city EMR classes. (No refs.) - M. Plessinger.

2776 *Berufe für behinderte Jugendliche (Occupations for handicapped youths)*. Second edition. Wiesbaden, Germany, Universum Verlagsanstalt, 1968, 367 p. (Price unknown).

Occupations most suited for the MR include repetitive work on assembly lines, service jobs (cooks, maids, and, in some cases, salespersons), and outdoor work in gardening and the building trades. Since training for some of these jobs usually takes longer, the employer must be willing to spend extra time to work with the trainees. On the positive side, it has been found that MR workers are more conscientious about performance and show a lower rate of absenteeism. During and after training, the MR should have an opportunity to continue their general education. If they desire, they should be able to reside in a special residence or be under supervision by a social worker. Before they are sent to a job, a complete evaluation of their abilities should be made and personal wishes considered.

It should be noted that the occupations and traineeships listed in this book apply only to the Federal Republic of Germany.
(237-item bibliog.) - S. L. Hammerley.

CONTENTS: Traineeships and Training Programs; Handicaps and Occupations; Occupational Guidance; Description of Occupations.

2777 ZAEZT, JAY L. *Occupational Activities Training Manual for Severely Retarded Adults*. Springfield, Illinois, Charles C. Thomas, 1969, 107 p. (Price unknown).

Programing used in the Occupational Activities Training Program to improve the functional skills of SMR children at Brandon Training School (Vermont) has been aimed at the development of self-help skills, physical coordination, attention span, and tolerance to simple tasks. The primary emphasis of the adult program is on the development of pre-vocational skills and preparation for employment in a sheltered workshop. The occupational activities described in this manual provide experiences in exercises, assemblies, and procedures. Descriptions of each pre-skill exercise and product assembly include lists of necessary materials, step-by-step procedures, and possible errors. Data on related programs, experiences in the development of this program, and case histories are included. These activities have been used successfully with institutionalized MRs with an IQ range of 15-30. This book would be of interest to institution superintendents, occupational therapists, and special educators. (16-item bibliog.) - J. K. Wyatt.

CONTENTS: The Outlook of the Occupational Activities Training Program; Related Programs in Preparation of Student Towards Occupational Activities; Procedure Experience with the Occupational Activities Training Program; Case Histories of Severely Retarded Students Who Were Enrolled in the Occupational Activities Program; Preskills for the Occupational Activities Program; Assemblies of the Occupational Activities Workshop; Introduction and Instructions to the Activities Checklist; Summary.

2778 PINECREST STATE SCHOOL. Validation of the Vocational Capacity Scale Utilizing Institutionalized Retardates. Final Report of Project RD-1619-P. Pineville, Louisiana, 1968, 129 p. Mimeographed.

An analysis of the validity of the Vocational Capacity Scale (VCS) with 366 institutionalized MRs (CA 15 to 30 yrs) revealed that the VCS maintained high levels of reliability and predictive validity and measured the factors of non-intellectual routine work, general ability, general health, and social maturity. The VCS was administered to all Ss twice, one year apart. It differentiated between the job levels of sheltered employed, competitively employed, and non-employed as well as among 5 related job levels within the sheltered-employed group. Normative data on the VCS for institutionalized MRs were different from data obtained in a community workshop setting. These findings indicate that the VCS can be used in the rehabilitation process to predict vocational potential and level of employment and to suggest areas in which training is needed. (72 refs.) - J. K. Wyatt.

2779 MEDICAL FOUNDATION. Post-School Vocational Adjustment of Educable Mentally Retarded Boys in Massachusetts. Mudd, Merle W., Melemed, Brina B., & Wechsler, Henry. (OEO Grant CG-9034). Boston, Massachusetts, 1968, 195 p. Mimeographed.

A study of the post-school vocational adjustment of EMR men who had attended public school special education classes in Massachusetts revealed that the majority had made a good vocational adjustment. Vocational adjustment was defined as the percentage of time S was employed full time since leaving school. Interviews were conducted with 549 EMR (IQ range 50-79) boys who had terminated attendance in special education classes during 1961 and 1962 and with 90% of their families. The interviews were designed to obtain a picture of current vocational adjustment and data on personal and environmental characteristics. About 1/2 of the Ss had been employed full time for 90% or more of the time since leaving school, and approximately 2/3 had been employed full time for 76% or more of the time. Although the Ss had been classified as EMR during their school years, the majority could not be distinguished from other individuals of the same age and social background after leaving

school. The longer a S had been in a special class for EMRs, the poorer his post-school vocational adjustment. Most schools had not provided specialized vocational training or preparation for work, and Ss who had obtained post-school special training exhibited poorer adjustment. Job hunting tended to be informal, and this type of job hunting tended to

be related to higher vocational adjustment. Higher social class membership was related to poorer vocational adjustment, a tendency to remain beyond age 16, and post-school training. The parents of higher social class Ss were less likely to expect their boys to find full-time employment upon terminating from school. (45-item bibliog.) - J. K. Wyatt.

TREATMENT AND TRAINING ASPECTS - THERAPY

2780 WEBB, RUTH C. Sensory-motor training of the profoundly retarded. *American Journal of Mental Deficiency*, 74(2):283-295, 1969.

Thirty-two PMR institutionalized children with encephalopathy were given sensory-motor training for 5 to 10 months. Training techniques to develop motor reactions to sensory stimulation are outlined. Clinical analysis of changes between pre- and post-treatment behavior tended to be more meaningful than the statistical comparison. This is attributed to the dissimilarity between measuring instruments used. A tentative theory underlying the application of these sensory-motor techniques to the profoundly retarded is presented as a point of departure for future theory formulations. (5 refs.)
Journal abstract.

Glenwood State Hospital and School
Glenwood, Iowa 51534

2781 FREDERICKS, H. D. BUD. Improved coordination in mongoloids. In: Washington, Institutions Department. *Proceedings of the Ninth Annual Research Meeting*. Olympia, Washington, *Research Report*, 2(2):68-71, 1969.

A behavior modification program was used to improve the coordination of 24 institutionalized mongoloid children (CA range 7-12 yrs). Behavior modification principles included shaping (from gross to finer movements), reverse chaining, and social reinforcement (hug, squeeze, kiss, "good", "very good", "wonderful"). Eight activities were developed, and each S was programed for 4 activities a day. Treatment, provided by college students, lasted for 20 minutes each day. Activities included string winding, board walking, maze

drawing, and a 6-phase pencil task. To permit a trend analysis for pre- and periodically-tested Ss and to control for constant testing effect, 1/2 the experimental and 1/2 the control Ss were pretested every 2 weeks and post-tested; the remaining Ss were post-tested only. Differences between periodically-tested and post-tested only groups were not significant. Mean post-test scores for the experimental group were significantly higher ($p=.05$) than those of the control group. A trend analysis on the scores of periodically-tested Ss revealed that the slope of the curve for experimental Ss was much sharper than the slope for control Ss. The behavior modification group demonstrated significant ($p=.05$) improvement in gross and fine motor coordination scores on the Lincoln-Oseretsky test. There were no significant differences between post-test scores and follow-up scores (3 mos after post-testing). (No refs.)
J. K. Wyatt.

2782 ISON, DAVID A., & NAUMANN, THEODOR F. Developmental training for the multiply handicapped. In: Washington, Institutions Department. *Proceedings of the Ninth Annual Research Meeting*. Olympia, Washington, *Research Report*, 2(2):72-77, 1969.

Changes in the environment of an institution and the application of specific training programs resulted in significant improvement ($p=.001$) in developmental skills for 35 multiply handicapped institutionalized PMRs and SMRs over an 8-month period. Positive social reinforcement was manipulated to decrease the number of bowel movements in non-verbal "crib" patients. The developmental training project then removed the Ss from their cribs and established a routine which would keep them out of bed. The program helped Ss to develop more diverse behaviors and greatly reduced self-abusive behaviors. Precise

training for self-feeding, communication, socialization, and occupation utilized a series of sequential steps and were based on very tight progress records. Comparison of project Ss with a comparable group of non-project Ss revealed significantly greater progress ($p=.025$) for project Ss over an 8-month period. These data indicate that relevant developmental experiences should be provided for multiply handicapped PMRs and SMRs. (No refs.) - J. K. Wyatt.

- 2783 PAUL, HOWARD A., & MARKOW, MICHAEL J.
Neurological organization exercises on retarded children with strabismus. *Training School Bulletin*, 66(2):66-71, 1969.

Three groups of retarded children were utilized to test the hypothesis that "an exercise program of the neurological organization type will reduce strabismus in retarded children and will result in an increase in performance." This increase in performance will be measurable on an intelligence scale. Group 1 consisted of 5 children with strabismus. Children in Group 2 (5 Ss) and Group 3 (10 Ss) had no strabismus. Groups 1 and 2 were given the experimental treatment of an exercise program of neurological organization type. Group 3 received a non-specific exercise program. After 6 months, the average mental growth of Group 1 was 6.6 months. There was a measurable correction of strabismus in this group. Groups 2 and 3 had 3.3 and 3.2 months mental growth. The effect on Group 1 was not permanent. A reduced number of Ss at completion of study made hypothesis testing impossible. (6 refs.) - *Journal abstract*.

American Institute for Mental Studies
Vineland, New Jersey 08360

- 2784 GRASSELLI, ANGELA, & MAGNONI, LILIANA.
Rilievi sui risultati ottenuti in quattro anni di trattamento rieducativo di bambini con p.c.i.--Confronto fra un gruppo di bambini trattati in famiglia e un gruppo di bambini trattati in centro ambulatoriale (Results of four years of work on re-educative treatment of cerebral palsied children--Comparison between a group of children treated at home and a group of children treated in an ambulatory center). *Neuropsichiatria Infantile*, 100-101 (May-June):465-480, 1969.

A 4-year observation of the fundamental motor functions of a group of 54 cerebral palsied children, of which 33 were trained at home and 21 in an ambulatory center, showed improvements in both cases, with a small, but real, advantage of the home training over

that received in the center. Each child was observed for a period of 1-2 years. The home training was performed by the children's mothers, who were instructed and guided by the Children Department Staff of the Istituto Neurologico "C. Besta". The training at the ambulatory center was performed by a physiotherapist. The proximity of the mother to the child during the day and the delegation of the responsibility of the training to the mother were factors contributing to the improvement shown by the home training group. (4-item bibliog.) - G. Pura.

Istituto Neurologico "C. Besta"
Milan, Italy

- 2785 MIHALOV, THELMA, MORALES, PABLO A.,
BADELL-RIBERA, ANGELES, & SWINYARD,
CHESTER. The management of incontinence in children with spina bifida and myelomeningocele. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 109-113.

A bladder management program for incontinent children with spina bifida and myelomeningocele should consist of scheduled voidings, regulated fluid intake, mechanical assistance of micturition, and the use of external appliances to keep the patient dry and/or hygienic. The Child's Urinal Bag is a new type of urinal for males which is comfortable, odorless, and inexpensive. It can be easily modified to meet a variety of needs. Urological evaluation is recommended during the neonatal period and at 6- to 12-month intervals, thereafter. Management of female patients involves the use of pads, waterproof pants, and scheduled voidings. Intractable incontinence in females can be treated by ileo-urostomy in some cases. (6 refs.)

J. K. Wyatt.

- 2786 TZIMAS, NICHOLAS A. Orthopedic care of the child with spina bifida. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 45-65.

Orthopedic services for children with spina bifida should be designed to help them become as independent as possible. Treatment may be

required to prevent the genesis of trophic ulcers and the development of deforming contractures and to correct deformities in the spine, hips, legs, and feet. To prevent trophic ulcers, all pressure must be either removed from vulnerable areas or must be redistributed over a wider area, with good skin hygiene maintained. Deforming contracture can be prevented: by not allowing the patient or his limbs to remain in the same position for long periods of time; by instituting passive range of motion in all joints, twice daily, and careful stretching of the contractures; and by using bracing in children over 12 months of age to maintain the limb segments in proper relation to the trunk and each other and enable standing and ambulation. Soft tissue operations to correct deformities include capsular and/or ligamentous and fascial release operations, tendon release or segmental resection operations, and tendon transfers. Bone operations include bone fusion procedures, osteotomy, and bone plastic operations. (26 refs.) - J. K. Wyatt.

- 2787 PEACH, WALTER, & HETTICK, DONNA. A speech stimulation program for institutionalized retardates. *Central Missouri Synthesis on Mental Retardation*, 1(1):1-7, 1968.

Individual speech instruction for institutionalized MRs (IQ 25-57; CA 7 to 15 yrs; 9 boys and 9 girls) resulted in improved speech and/or behavior. A pre-behavioral report stated each MR's receptive language, expressive language, and behavior. Training was individualized and typically included identifying a doll's clothing, 3 colors, a circle, and a square. Improvement in behavior and/or speech was noted in 12 Ss, although maturation or individual attention may have influenced changes. (4 refs.) - M. Plessinger.

Central Missouri State College
Warrensburg, Missouri 64093

- 2788 MONACO, THERESA M., PEACH, WALTER, BLANTON, RICHARD S., & LOOMIS, DORIS. Pilot study: Self care program for severely retarded girls. *Central Missouri Synthesis on Mental Retardation*, 1(1):8-20, 1968.

Institutionalized SMR girls (CA 114-184 mos; IQ < 40) who needed complete or partial help to dress responded to training on dressing themselves. Each girl's background was tabulated, and the attendants' before and after evaluation was obtained. The Ss were given 30 minutes of individualized training for 16 days during which time the Ss dressed and undressed themselves, with verbal and demonstration

cues given as needed. Reinforcement was either verbal, a pat on the face or candy for appropriate responses to verbal assists or no assists. The number of verbal and demonstration assists was tabulated at each session. Five Ss showed definite improvement. Conclusions were: time needed to improve dressing varies; demonstration aids dressing training; trainees do respond to verbal and demonstration assists; and SMR respond to controlled training when emphasis is on positive results. (2 refs.) - M. Plessinger.

Central Missouri State College
Warrensburg, Missouri 64093

- 2789 BRACKEY, LYNN. How our flowers grow. *Challenge* (Newsletter of Project on Recreation and Fitness for the Mentally Retarded), 4(3):3, 1969.

A charm school for TMR girls was successful in teaching acceptable social behavior and good grooming through instruction and practical experience. Learning was reinforced by incentive awards, regular progress assessments, and the cooperation of parents. (No refs.) - E. F. MacGregor.

Duval County Recreation Department
Jacksonville, Florida

- 2790 HUMES, CHARLES W., JR., ADAMCZYK, JOHN S., & MYCO, ROBERT W. A school study of group counseling with educable retarded adolescents. *American Journal of Mental Deficiency*, 74(2):191-195, 1969.

This is an experimental study designed to differentiate outcomes among treatment combination groups with counseling and no-counseling as the independent variables. Ss were 28 EMRs, ages 13 to 17 and IQs 53 to 77. A 2 X 2 design was used with 2 levels of experimenter variables and 2 of treatment conditions. The groups received 12 hours of counseling or no-counseling. Post-testing was done with a variety of instruments measuring classroom behavior, personal-social factors, self-concept, interpersonal relationships, and tested for .05 significance. Results indicated that counseling can be effective with this population in some behavioral areas and with certain assessment instruments. (12 refs.)
Journal abstract.

Westfield Public Schools
Westfield, Massachusetts

2791 AYER, M. JANE, & BUTLER, ALFRED J.

Client-Counselor Communication and Interaction in Counseling with the Mentally Retarded. (Final Report RD 1798-P). Madison, Wisconsin, University of Wisconsin, 1969, 179 p.

This study described and measured the counseling interaction with moderate to borderline MR adolescents and young adults in a residential school. The project consisted of 6 related studies which were completed over a 3-year period and utilized 4 approaches in the measurement of the counseling process. The study showed that it is possible to assess client verbalization and that the clients recognized, verbalized, and perceived a

significant number of problems of a cognitive nature. The affective aspects of counselor behavior appeared to reinforce expression of the cognitive elements of client problems. Client-counselor communication stressed exchange of information, the content of which was related to language and personality characteristics of the client. Individual differences among counselors in their approach and method were demonstrated, and the amount of client-counselor interaction was related to age and intelligence of the MR. The Verbal Expressivity Scale (VES) was designed as a measure of the client's ability to communicate relevantly. Further research is needed to clarify the usefulness of the VES as a diagnostic tool and as a measurement of the counseling process. (147 refs.) - S. Half.

PROGRAMMATIC ASPECTS - PLANNING AND LEGISLATIVE

2792 INTERSTATE CLEARING HOUSE ON MENTAL HEALTH.

Action in the States in the Fields of Mental Health, Mental Retardation, and Related Areas: A Report on 1964-1968 Financial, Legal and Administrative Developments in the States' Mental Health Programs. Chicago, Illinois, 1969, 108 p. \$5.00.

Recent developments in the United States in the area of services for the MR have emphasized the expansion of schools and institutions, the replacement of custodial care with a training and habilitation approach, an increase in educational and vocational services, the provision of diagnostic and screening services in institutions, and mandatory special education in public schools. Day-care programs, many of which are state-supported, are increasing, and regional diagnostic centers have been established in some states. Additional diagnostic, evaluation, and outpatient services are becoming available. School programs for EMRs are available in

most states, and many of these now include TMRs. At least 40 states now have laws which require the testing of neonates for PKU and other metabolic disorders. Specific descriptions of MR and special education developments in each state are included. There is a state-by-state breakdown of capital outlay for public institutions as well as descriptions of patient care and community mental health programs. (No refs.) - J. K. Wyatt.

CONTENTS: Summary of Recent Developments - 1964-1965; Summary of Recent Developments - 1966-1967; Community Mental Health Services; Research and Training; Patient Care; Mental Retardation and Special Education; Foster Care; Volunteers; Organization and Legislation; Committees and Commissions; Effect of Planning on Program Developments; Role of Mental Health in Comprehensive Health Planning on Program Developments; Role of Mental Health in Comprehensive Health Planning under P. L. 89-749.

2793 WHITE, WESLEY D. *Planning and Programming for the Retarded: Yesterday, Today and Tomorrow*. New York, New York, National Association for Retarded Children. 1969, 8 p.

The MR should be afforded every opportunity to grow and develop to their maximum potential, and this can best be accomplished in a home-like environment where each child is exposed to enriching experiences. MRs must be recognized and accepted as human beings and individuals entitled to a happy, full, and rewarding life. The philosophy of yester-year encompassed concepts, attitudes, and approaches toward the MR which led to their dehumanization. New ideas, meaningful goals, and significant programs must be based on current research and knowledge. Programs are needed to motivate and stimulate the MR. Staff should utilize every available skill and technique to enhance the MR's social and mental development. The most suitable and desirable placement program is a family-like group in a small house-like unit where stress situations will be minimized, dependency lessened, wholesome interpersonal relationships established, and healthy interaction with adult and peer groups realized. It is necessary that the public be re-educated and the communities and families be more understanding and accepting of MR. There will continue to be a need for institutional placement to care properly for those children with special problems who are unable to live at home or be cared for in community residential centers. Improvement of programs for the MR can be brought about by smaller, more individualized living units, better quality of supervision, more concerned staff, more modern and positive attitudes in the administrative structure, and in-service training programs for aides and attendants permitting them more flexibility and less rigidity. Failure to obtain additional financial assistance is not always a detriment in providing program improvement in institutions. Present day rationale should be to de-emphasize the sheltering of the MR from society and assist them in assuming their role as participating members of society. Major steps should be taken toward integrating the MR into the mainstream of society. (No refs.) - S. Half.

2794 DONDORP, D. W., JR. De methodiek in het sociaal pedagogisch werk (Methodology of social and pedagogical work). *Tijdschrift voor Zwaksinnigheid en Zwaksinnigenzorg*, 5(1):14-25, 1968.

A new technique which is currently being developed in The Netherlands for the post-schooling care of the MR combines educational

and social work methods in an effort to make the MR's adjustment to society more complete. Many MRs can be integrated into normal society through proper counseling, appropriate job placement, and the provision of opportunities for social contacts. This can be done on a one-to-one basis as well as in groups. At present, no definitive system has been established; such a system, however, may limit rather than expand the possibilities for the MR in this area. (No refs.)

S. L. Hamersley.

No address.

2795 DICKE, W. Eingliederungshilfe für behinderte Kinder und Jugendliche in der BRD (Aid in rehabilitation of handicapped children and youth in the Federal Republic of Germany). *Die Rehabilitation*, 7(3):134-136, 1968.

To improve the effectiveness of present rehabilitation work with the physically handicapped, the number of early diagnostic and treatment centers must be increased, more special kindergartens outside of the metropolitan areas must be opened, and the number of sheltered workshops must be expanded. Government and private institutions should cooperate in achieving these goals. (No refs.) - S. L. Hamersley.

Alexanderstrasse 23,
Stuttgart 1, West Germany D 7000

2796 GROSS, HERBERT. Eingliederungshilfe für behinderte Kinder und Jugendliche in der BRD (Aid in rehabilitation of handicapped children and youth in the Federal Republic of Germany). *Die Rehabilitation*, 7(3):136-146, 1968.

A course of action to improve the rehabilitation of physically and mentally handicapped children in the Federal Republic of Germany should include: a census of the handicapped and types of disabilities, improvement of existing facilities and creation of new ones, and increased social work with the families of the handicapped. At present the number of handicapped children in the Federal Republic and the exact nature of their disabilities is not known. Financial aid for research on this and many other projects is necessary. Direct financial aid to the handicapped who are unemployable and to their families should be considered. Not enough attention is being given to the emotional and social problems created by the presence of the handicapped

child in the family. The total load of rehabilitation work, which could be coordinated at the federal level, should be distributed among the federal and state governments and private agencies. (No refs.)
S. L. Hamerley.

Kirchfeldstrasse 149
Dusseldorf, West Germany D 4000

2797 SECRETARY'S COMMITTEE ON MENTAL RETARDATION. National center on educational media and materials for the handicapped. *Programs for the Handicapped*, Newsletter of the Secretary's Committee on Mental Retardation, U. S. Department of Health, Education, and Welfare. 69-12 (September 30), 1969, 5 p.

Public Law 91-61, passed in 1969, provides for a National Center on Educational Media and Materials for the Handicapped to use new educational technology in programs to educate the handicapped. Since teachers are in demand and only 40% of the 5 million handicapped children are being served, the Center will centralize research, production, and distribution of instructional media and materials. The Center will link together various

instructional materials and media centers, research projects on curriculum, and film distribution centers. The Bureau of Education for the Handicapped in the U. S. Office of Education will administer the Center which will receive \$12.5 million in 1971, \$15 million in 1972, and \$20 million each year thereafter. (No refs.) - M. Plessinger.

2798 VAN DER KELEN, R. Statuut buitengewoon onderwijs nakend (Are the laws concerning special education insufficient)? *Amentia*, (December):12-14, 1968.

In the realization that the law is no cure-all for educational problems, these recommendations are made for the revision of the present Belgian laws: more carefully delineated admission criteria when admitting children to special education schools (not all handicapped children belong there), higher educational requirements for teachers of the handicapped, construction of more facilities, and the provision of free education. (No refs.) - S. L. Hamerley.

No address.

PROGRAMMATIC ASPECTS - COMMUNITY

2799 SUSSER, M. W. Community approaches to rehabilitation in mental subnormality. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 26, p. 203-208.

Services for the MR in Salford (England) emphasize socialization, which is defined as rehabilitation. A register of all MRs is kept so that community services can be planned for their entire life cycle, for all degrees of handicap, and for the study of the dynamics of the MR population. The population toward whom rehabilitation services are aimed are the 1/3 to 1/4 of the mildly MR who fail to make adequate social adjustments. Less than 1/3 of this group is composed of individuals with learning handicaps due to organic brain disorders, sensory loss, emotional disorders, or chronic diseases of childhood. The remaining 2/3 are clinically and socially homogeneous, have no detectable handicaps, are primarily drawn from the most

disadvantaged social groups, and tend to make intellectual gains in young adulthood. Day care programs and adult centers for individuals with clinically recognizable MR provide special education for individual handicaps, specific training to help the individual remain in society, sheltered occupational niches, a milieu which enhances the development of socialization, and personal and family support. Although individuals with cultural MR require some of these same services as well as prolonged schooling, the main challenge which they present is one of prevention through social policy. When individuals have been exposed to cultural MR and family deprivation, socialization becomes distorted to a point where they are not capable of sustained normal interaction. The prevention of distorted socialization depends on finding ways to help disordered families as soon as they are identified, experimenting with and evaluating new types of residential facilities, and avoiding long-term confinement in social institutions. (33 refs.) - J. K. Wyatt.

2800 BABOW, IRVING, & JOHNSON, ALONZA C.
Staff attitudes in a mental hospital which established a mental retardation unit. *American Journal of Mental Deficiency*, 74(1): 116-124, 1969.

To study the attitudes of the treatment staff at a state mental hospital which established a MR unit, a questionnaire was constructed which included attitudes toward MR, the F scale on authoritarianism, Sroles's anomia scale, and the Strauss-Schatzman scales on psychiatric treatment ideology. Of 1,170 questionnaires distributed, 760 usable ones were returned. Intercorrelations were made between the MR orientation and the 5 scales. Two major factors were found in the MR orientation. An O analysis of respondents' attitude profiles yielded 5 clusters. These were analyzed and the implications for patient care and hospital organization were discussed (11 refs.) - *Journal abstract*.

Napa State Hospital
Imola, California 94558

2801 GREATER OMAHA ASSOCIATION FOR RETARDED CHILDREN. *The Initiation and Development of a Comprehensive, County-Wide System of Services for the Mentally Retarded of Douglas County: A Planning Report to the Douglas County Board of Commissioners*. Menolascino, Frank, J., Clark, Robert L., & Wolfensberger, Wolf. Omaha, Nebraska, 1968, Volume 1, 82 p.

A plan for the development of a comprehensive, county-wide system of community services for MRs of Douglas County (Nebraska) includes a proposal for the establishment of a County Department of MR which would provide essential services for MRs not now provided by generic service agencies. Among the services to be developed are developmental day-care centers, crisis assistance units, family evaluation and guidance services, recreational programs, residential care, and sheltered workshops. There are probably 12,000 MRs in the county; however, only 1/6 have been identified. The plan calls for the establishment of small local residential units so that 634 county citizens institutionalized in a State home can be returned to their home county. During the first year of the plan's funding, a central office and administrative and planning staff were developed, operation of the County Vocational Services Center and the Benson Development Center was begun, and the first workshop in Nebraska which is exclusively for the vocational training of MRs was established. (No refs.) - J. K. Wyatt.

2802 WESTERN INTERSTATE COMMISSION FOR HIGHER EDUCATION. *Four Corners Mental Retardation Project: The Training and Use of Indigenous Aides in a Sparsely Populated, Economically Depressed Region: Final Report*. Boulder, Colorado, 1969, 87 p.

The goals of the Four Corners MR Project were to identify the needs of MRs in the area and to identify and enhance available services for the handicapped. The Four Corners Area, a poverty area, includes adjoining sections of Utah, Colorado, Arizona, and New Mexico; 8.2% of its residents have a Spanish surname, and 48.3% are Indian. The project has developed a *Directory of Services Available to the MR in the Four Corners Area*, trained 10 indigenous staff members in the basic aspects of MR and placed them in several communities, surveyed agencies to identify the number of handicapped persons in the area, stimulated social agencies to provide additional services for MRs, and coordinated preliminary inquiries into the development of a network of sheltered workshops among the 4 states. The survey identified 1989 handicapped persons (869 MRs) of whom approximately 900 (45.3%) were Indians. Breakdown of handicap by age revealed a need for the expansion of classes for EMR and TMR children, and a great need for an institution specifically for the PMR. Comprehensive services in rural areas and small communities are needed; specifically, physical, mental health, and social work services; work-training programs; transportation; and recreation. Since persons in this area have little knowledge about MR or about programs for MRs, a public awareness program is needed. Immediate and major emphasis should be placed on the use of additional indigenous personnel in the helping services with continued emphasis on the provision of cross-cultural experiences in the training of multi-cultural professionals. (5 refs.)
J. K. Wyatt.

CONTENTS: Field Work; Survey of Handicapped Persons in the Four Corners Area; Services Needed in the Four Corners Area; Program Development; Recommendations.

2803 CALDWELL, BETTYE M., RICHMOND, JULIUS B., HONIG, ALICE S., MOLDOVAN, STANLEY E., MOZELL, CHARLENE, & KAWASH, MARY B. A day-care program for disadvantaged infants and young children--Observations after one year. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, p. 103-115.

The aim of a day-care intervention program for culturally disadvantaged children under 3

years of age was to provide an atmosphere in which the children can thrive. The program used an individualized curriculum oriented toward the development of the personal-social, cognitive, and breadth of experience aspects of each child. The adult-child ratio was 1:4, and the staff was multidisciplinary. The distribution of parent occupational and educational levels was bimodal. Of 23 separate families in the sample, the education range of the fathers was from sixth grade to a doctor of medicine degree with the missing fathers of the 11 fatherless families having minimal educations. The modal income for 11 families was \$250/month, 4 families had incomes in the \$250-\$450 range; and the income of 8 families exceeded \$450/month. After 7.5 months in this program, the mean developmental quotient of the children had increased significantly ($p < .01$) from 104.5 to 110.1. The children in this program received a good foundation for the establishment of a basic sense of trust in people and the environment, and they were encouraged to use their own senses and experiences to understand and master their worlds. (11 refs.) - J. K. Wyatt.

2804 VULPE, SHIRLEY GERMAN. *Home Care and Management of the Mentally Retarded Child*. Toronto, Ontario, Canada, National Institute on Mental Retardation of the Canadian Association for the Mentally Retarded, 1969, 165 p. (Price unknown).

The goals of home-care programs for families with an MR child are to provide support, programs, counseling, and instruction in special techniques which will reduce the effects of the handicaps on the child, help the parents and child to function at their maximum ability levels, prevent later maladaptation due to early mismanagement, and reduce parent frustration. Individual treatment programs should be based on the findings of comprehensive assessments of the child, his family, and his environment. A behavioral and developmental assessment battery designed to determine whether an MR child meets the criteria for a home program which uses occupational therapy techniques and procedures is described in detail. The battery can be used with children from 3 months to 6 years of age, and can be adapted to the special needs of the child and test administrator. Activities, equipment, and techniques which have been useful in home-care programs and methods of designing and advancing programs are described. This manual would be of interest to occupational therapists, social workers,

educators, psychologists, and nurses. (20-item bibliog.; 53 refs.) - J. K. Wyatt.

CONTENTS: Introduction; Selection of Parents and Children for Home Care Services; Setting and Staff; Assessment for Treatment; Methods of Planning Programs; Specific Techniques; Progression of Programs and Activities; Treatment Apparatus and Equipment; Sample Programs.

2805 O'HARA, BRIAN, PATTULLO, ANN, & GRANT, JIM. *Child Study Clinics in Washington State*. Washington. Institutions Department. *Proceedings of the Ninth Annual Research Meeting*. Olympia, Washington, Research Report, 2(2): 53-56, 1969.

Although the major focus of the diagnostic services provided by the 15 Child Study Clinics in Washington is on the needs of MRs, current programs are oriented toward the diagnosis and treatment of childhood handicaps. The work of 3 full-time and 12 local clinics is coordinated by a consulting team (pediatrician, social worker, and public health nurse) from the University of Washington. This team fosters communication, provides in-service seminars, and arranges orientation programs for local professionals. Of 671 new patients diagnosed in 1967-1968, 58% had IQs below 75. In spite of the availability of biochemical screening, etiology was unknown for over 1/2 of the cases of MR seen in 2 regional and 10 local clinics. The clinic populations evidenced a significant number of physical and emotional handicaps. The distribution of diagnostic clinics conforms to the diagnostic and service areas recommended in the 1966 State Mental Retardation Facilities Construction Plan. (4 refs.) - J. K. Wyatt.

2806 CANADIAN ASSOCIATION FOR RETARDED CHILDREN. *Guardianship for the Mentally Retarded*. Toronto, Ontario, Canada, No date, 11 p.

Parents need advice when they are making lifetime financial plans for their MR child. The Canadian Association for Retarded Children has established a Guardianship Committee to aid and stimulate parents in the personal planning for the MR and to explore more adequately and develop data for those individuals and professionals concerned with the various aspects of this vital problem. The Canadian Association finds that the prime problems involved in developing a meaningful guide are the diversity of the needs of the MR and their families and the variations of the laws and practices of the provinces. The

Guardianship Plan should precisely outline the kind of care the family wishes their retarded offspring to have when the parents expire or become incapacitated to the extent that they are no longer able to provide adequate care and supervision. Many purposeful and legal ways for families to assure financial protection for their MR child throughout his life span can be arranged--a trust fund, life insurance, securities, real estate, stocks, bonds, jewelry, and other assets. The most significant factor is that the parents assume responsibility and make sure that

their will gives specific and precise directions in regard to the allocation of all monies in order to guarantee financial security for their child. Parents should name a guardian who will be responsible for the management of all inherited assets and the child's care, supervision, education, and training. If parents wish, they can prearrange placement of the child in a residential setting; however, this agreement with the director of a specific facility should be clearly identified in the will. (No refs.)
S. Half.

PROGRAMMATIC ASPECTS - RESIDENTIAL

2807 Residential care for the mentally retarded. *Programs for the Handicapped*, (Newsletter of the Secretary's Committee on Mental Retardation), 68-11, October 18, 1968, 20 p.

Institutions for MR are overcrowded, and the care is generally poor quality since states usually lack appropriate financing; therefore, federal funds have been made available. The Department of Health, Education, and Welfare provides direct funds and indirect aid to institutions. The Rehabilitation Services Administration (RSA) gives grants-in-aid to states to develop rehabilitation and vocational programs; the Hospital Improvement Program also funds state institution programs. Other RSA programs include: Summer Work Experience and Training (SWEAT) in MR for high school seniors; construction of the Community Facilities Program; University-Affiliated Facilities Construction Program; and Hospital In-Service Training. Other agencies which provide funding and programing for the MR include: The Children's Bureau which administers federal grant programs for child welfare services; the Medical Services Administration; the Administration on Aging which administers the foster grandparents (4,000) program; the National Institute of Child Health and Human Development which administers construction programs of research centers; the National Institute of Mental Health which collects and publishes data on the MR in public and private institutions; the Office of Education which provides assistance

for library services and construction and education for the MR in institutions; the Social Security Administration which provides benefits for the MR; and the Surplus Property Program which makes land available for MR hospitals and schools. Since 1966, military MR dependents are eligible for services and care. (12-item bibliog.) - M. Flessinger.

2808 AMERICAN HOSPITAL ASSOCIATION. *Developing Policies and Procedures for Long-Term Care Institutions*. Chicago, Illinois, 1968, 56 p. \$1.75.

A carefully written policies and procedures manual is an important guide to decision making and efficient management and benefits the entire hospital staff, the patients, and the general public. Policies should be made by the governing body of the institution in conjunction with the recommendations of an advisory group. They provide directions for action which may be amplified by the medical or administrative staff to cover day-to-day needs. Procedures provide step-by-step instructions on how policies are to be activated and should be developed by the people responsible for performing them. A policy and procedure manual for a long-term care institution should contain a brief history and background of the institution, its goals, definition of terms, and the policies and procedures. Recommendations for the development

and preparation of a manual are included. Illustrations of general administrative, service, and departmental policies and procedures in relation to communication with employees, vacations, admission of patients, physician's services, nursing care plans, and patient's dining room are described in detail. This book should be of interest to hospital administrators. (82-item bibliog.)

J. K. Wyatt.

CONTENTS: Introduction; Developing and Writing Policies and Procedures; and Contents of a Policy and Procedure Manual.

2809 CLELAND, CHARLES C., & PATTON, WILLIAM F. Sustained versus interrupted institutionalization. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 24, p. 192-198.

The performance of 88 institutionalized MRs on academic achievement tests administered at the beginning of and following summer recess was evaluated. Fifty-three Ss (mean CA 14.2 years; mean IQ 62.1) received furloughs of 30 days or more, and 35 Ss (mean CA 13.9 yrs; mean IQ 61) were in continuous residence. Forms B and C of the Gray-Votaw-Rogers General Achievement Test were used to measure academic performance level. There were no significant differences between the academic achievement scores of tenure-interrupted and noninterrupted groups. The tenure-interrupted group evidenced the only decrement in performance. These findings may be due to differential types of reinforcement experienced by the Ss during the periods between pre- and post-testing. Sustained institutionalization may be an important factor in the rehabilitation of MRs. (14 refs.)

J. K. Wyatt.

2810 ABELSON, ROBERT B., & *PAYNE, DAN. Regional data collection in state institutions for the retarded: Reliability of attendant ratings. *American Journal of Mental Deficiency*, 73(5):739-744, 1969.

The reliability of ward attendants to provide ratings on patient characteristics and behavior was examined. The data collection form consisted of 78 multiple-choice items with 2 to 8 choices/item on topics which ranged from patient's sex to questions concerning social and emotional behaviors. The agreement between attendant ratings and ratings by professional staff was obtained by having each group rate the same 285 residents. Test-retest data were obtained on 646

patients; and the similarity between morning and evening shifts was compared for 287 patients. Percent agreement was computed in each case. Items not rated by one or both of the raters were eliminated. In each study, the median percent agreement was over 80%. Generally, objective items with few alternatives had the highest reliability. Items requiring memory over a long period of time had low reliabilities as did items requiring judgments about social and emotional behaviors. (5 refs.) - J. M. Gardner.

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Boulder, Colorado 80302

2811 AMERICAN PSYCHIATRIC ASSOCIATION. *Motivation for the Mentally Retarded: A Report from Fairview State Hospital, Costa Mesa, California*. Washington, D. C., Remotivation Project. No date, 12 p.

Motivation techniques were used in a pilot study at Fairview State Hospital (California) to help MRs learn to relate to their surroundings in a useful, meaningful way and to enable as many as possible to leave the hospital and receive "Home Care". The 8 boys (some with IQs between 12 and 20) who participated in the project were from a maximum security ward, were diagnosed as psychotic MR, SMR, moderate MR, or mild MR, and exhibited self abusive and antisocial behavior. They participated in group experiences led by an attendant-leader who made each S feel accepted and wanted, created a bridge to reality for each S, taught the Ss to share the world we live in and appreciate the work of the world, and made them feel appreciated. Although some Ss did not speak and were considered ineducable prior to participation in the project, they were, after 3 months, actively participating in group experiences and accepting instruction, and several Ss learned the alphabet. As their social habits improved and good relations with their friends and surroundings developed, the Ss became easier to manage. The successful use of motivation techniques requires program continuity, the use of simple and easily understood poetry or prose, and respect and interest by the attendant-leader in and out of group sessions. (No refs.) - J. K. Wyatt.

2812 SHELLHAAS, MAX D., & NIHIRA, KAZUO.

Factor analysis of reasons retardates are referred to an institution. *American Journal of Mental Deficiency*, 74(2):171-179, 1969.

A replication of Maney, Pace, and Morrison's principal component analysis of the needs for institutionalization was undertaken in a Midwestern institution for the MR. The present analysis did not yield a factor structure similar to theirs. Lack of agreement between the 2 studies was tentatively attributed to the heterogeneity of the institutional population in the present study. A rotated factor matrix resulted in 10 factors reflecting specific reasons retardates were institutionalized. These factors suggested the 3 major foci for programs designed to rehabilitate the MR mentioned by Maney *et al.*, and an additional area of program development that would be directed toward changing the basic social environment of the retarded. (4 refs.) *Journal abstract.*

Parsons State Hospital and Training Center
Parsons, Kansas 67357

2813 LYON, RICHARD, & BLAND, WAYNE. The

transfer of adult mental retardates from a state hospital to nursing homes. *Mental Retardation/MR*, 7(5):31-36, 1969.

This paper is an attempt to evaluate a 15-month-old program of placing hospitalized MR adult patients in nursing homes. The primary concern of the paper was to answer the question "Was placement in a nursing home 'better' for older MR patients than remaining in a state hospital?" The study consisted of obtaining general information on 94 patients and studying intensively (interviews and questionnaires) 25 patients in 14 nursing homes. It was possible to conclude that the patients placed were generally functioning at a much higher level than they had functioned in the hospital. The patients received "better" care, many were encouraged to help with nursing home tasks, and some were paid for chores performed. A high percentage were considered to be "happier" and more satisfied with the nursing home than they had been at the state hospital. (4 refs.) - *Journal abstract.*

Central State Hospital
Milledgeville, Georgia 31061

2814 MACKAY, HARRY A., & SIDMAN, MURRAY. Institutionalizing the mentally retarded in an institutional environment. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 21, p. 164-169.

Reinforcement techniques and programmed instruction were used to change a basically destructive "home" environment in an institution for the MR to one which supported constructive growth. The Ss were 71 institutionalized SMR boys (CA 6-20 yrs) who: required continuous supervision, and engaged in an aimless, stereotyped, and aggressive behavior; had to be spoon fed; exhibited minimal, poorly articulated verbal behavior; and were either partially or completely unclothed. The working staff of the institution was trained to use effective teaching methods and to relieve their physical burden by training the children as helpers. A token economy was established and used to elicit new and useful kinds of behavior. Laboratory-derived methods were used in a program of academic training. The techniques of programmed learning were used to specify the component behaviors of complex and terminal performances. Systematic procedures were developed to communicate with the children. In addition to caring for their own needs, many children learned to take complete care of the dining room, laundry, and bedroom, and some learned to toilet less-capable children. While this program did not increase intelligence quotients, it provided a practical means of teaching the children prerequisite skills which will permit a more valid assessment of their behavioral potential. (6 refs.) - J. K. Wyatt.

2815 BRONFENBRENNER, URIE. Institutional approaches to cultural deprivation--American and Soviet. In: Jervis, Charles A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 12, p. 93-102.

The major differences between American and Soviet preschool programs are that the Russians have provided new buildings with permanent equipment while such special facilities have not yet been provided by Americans and that Soviet programs neither reach into

the home nor use parents as volunteers. Soviet programs also serve a wider age range, and priority of admission is given to children from families where one parent is absent or where parents work on different shifts. The long-range goals of Soviet preschool training are to provide an early experience in collective living and to expose the child to a regime which is based on a series of reinforcement schedules. Emphasis is placed on sharing, cooperative activity, and collective activity. At the school-age level, the principle of mutual help is widely applied, and the child's "collective" becomes the primary disciplinary agent. Aspects of cultural deprivation highlighted by Soviet methods of collective child-rearing are that the culturally deprived child may suffer from an inadequate understanding of and commitment to the values of his society as well as from intellectual deficiencies and that the atmosphere of the child's peer group may hold the key to the development of this understanding and commitment. These factors have not yet been considered in American compensatory education programs. (No refs.) - J. K. Wyatt.

2816 LARSON, ELIZABETH RAE. Rainier School, Department of Institutions, State of Washington Hospital Improvement Project, Unit X: Establishing functional self-help skills

in severely and profoundly retarded adolescents and adults. In: Washington. Institutions Department. *Proceedings of the Ninth Annual Research Meeting*. Olympia, Washington, Research Report, 2(2):60-66, 1969.

The interdependent goals of the Hospital Improvement Project (Washington) were staff development, resident treatment and training, and program development. Personnel training was planned to develop non-professional staff members who could assess deficit and growth and design and institute behavior modification treatment techniques. Professional consultant services were provided. Most of the 96 SMR and PMR residents (CA range 8-40 yrs) demonstrated severe behavior problems. General treatment goals were aimed at developing independence in eating and toileting and reducing aggressive and destructive behaviors. The 70 project Ss made fairly consistent gains in feeding, toilet training, and tooth brushing and considerably less progress (negative in some cases) in grooming and dressing. Twenty-four control group Ss showed a slight loss in skills in all these areas. Analysis of communication deficits revealed that 15.6% of the Ss were non-verbal, 45.8% were non-verbal but vocal, and 38.6% demonstrated some verbal abilities. The results of this program suggest that non-professional staff members can be trained to meet the needs of MRs. (No refs.) - J. K. Wyatt.

PROGRAMMATIC ASPECTS - RECREATIONAL

2817 OLIVER, JAMES N. Recreation for the severely handicapped. In: Jervis, George A., ed. *Expanding Concepts in Mental Retardation: A Symposium*. Springfield, Illinois, Charles C. Thomas, 1968, Chapter 30, p. 232-237.

The recreational therapy department at St. Margaret's Hospital (Birmingham, England) has an ambitious program which successfully provides recreation for a large proportion of the 1,746 MR patients. The principal annual recreation projects are a concert and a pageant, and recreation time is mainly used to

practice for them. Patients are grouped by ability rather than mental age, and although the main emphasis is on helping older patients, children are included. Very low-grade, physically unfit patients with limited capacities participate as they are able, and their inclusion gives them feelings of belonging and accomplishment. Practices are progressive, and larger numbers of patients are involved as the time for the activity nears. The patients make costumes and scenery, and their involvement in these recreation projects over a period of many months has had far-reaching effects. (8 refs.) - J. K. Wyatt

2818 CARR, ADELL C. A therapeutic recreational program for children with spina bifida and myelomeningocele. In: New York University Spina Bifida Study Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 131-133.

A recreational program for children with spina bifida and myelomeningocele should capitalize on strengths, complement and supplement the training program, and provide the children with feelings of attainment and pleasure. The facilities and program should be designed to accommodate wheelchairs. The program should emphasize bi-manual activities and should include activities in a standing position for children who can stand. It should contain group activities, crafts, physical activities, free play, motor skill activities, and excursions. The recreation therapist should be aware of the emotional and behavioral characteristics of the spina bifida child and should focus on goals which will aid individual treatment. (No refs.)

J. K. Wyatt.

2819 AMERICAN ASSOCIATION FOR HEALTH, PHYSICAL EDUCATION AND RECREATION, & NATIONAL RECREATION AND PARK ASSOCIATION. *Physical Education and Recreation for Handicapped Children. Proceedings of a Study Conference on Research and Demonstration Needs*. Washington, D. C., 1969, 81 p.

The need for research in physical education and the creation of these programs were discussed, with the discussion centering on the areas of institutions, disciplines, disabilities, and functions as they pertain to handicapped children. Objectives included elimination of inappropriate use of federal funds, establishment of priorities, provision of a framework for evaluation, identification of basic research topics and qualified researchers, examination of the role of various agencies and present systems of disseminating research, and promotion of team work. Major statements were issued by the conference participants on the development of comprehensive dissemination procedures, the stimulation of relevant projects, the encouragement of better coordination between agencies, the analysis of the skills needed for participation in recreation, and the increase of research on learning by means of physical activities. (85-item bibliog.; 47 refs.) - V. G. Votano.

CONTENTS: Purposes and Objectives; Panel Presentation; Keynote Address; Recommendations.

2820 THOMPSON, MORTON. The status of recreation for the handicapped as related to community and voluntary agencies. In: American Association for Health, Physical Education and Recreation, & National Recreation and Park Association. *Physical Education and Recreation for Handicapped Children. Proceedings of a Study Conference on Research and Demonstration Needs*. Washington, D. C., 1969, p. 25-29.

Questionnaires were sent to 427 recreation departments to inquire into their programs for the handicapped and MR, and 202 responded. Forty percent of these communities segregated the handicapped, 2/3 of the programs were conducted in cooperation with other community agencies, and 1/3 were conducted in association with health agencies. Ninety-one percent of the recreation departments provided some programs for the handicapped. Staff members included 54 directors, 87 supervisors, 193 leaders, 48 part-time workers, and 1,100 volunteers. Ninety percent of the funds were provided by taxes in 56% of the communities; however, 13% of the agencies received no tax money. Program activities included arts and crafts, games, picnics, music, drama, carnivals, and hiking. Facilities included playgrounds, swimming pools, recreation centers, and day camps. Those communities which lacked recreational facilities for the handicapped gave as reasons poor budgeting, transportation expense, lack of staff members, insurance problems, and higher costs of such programs. Improved public relations, new studies, parent militancy, research, elimination of architectural barriers, and better staff training may be helpful in establishing new programs. (No refs.) - V. G. Votano.

2821 O'MORROW, GERALD S. The status of recreation for handicapped children in institutions. In: American Association for Health, Physical Education and Recreation, & National Recreation and Park Association. *Physical Education and Recreation for Handicapped Children. Proceedings of a Study Conference on Research and Demonstration Needs*. Washington, D. C., 1969, p. 29-35.

Because of lack of funds, staff shortages, and unconcern for patients, there are only very limited recreation programs for individuals in institutions. Learning and using skills are the only programs now operating. Recreation activities help the handicapped increase growth and develop a better self image, reduce isolation, redirect attention, establish constructive attitudes, reduce aggression, stimulate interest, and prepare the child for adult roles. Principles of the

leaders of these activities should include accepting the participant as a human being, involving the child quickly, starting at his existing level, being aware of the child's health, and having a positive attitude. Recreational counseling is needed both for outpatients and inpatients to enhance the capacity for social function. Counseling helps the child to explore his recreational needs and aids him in locating, using, and identifying community resources. The institutionalized child needs to participate in community recreational activities in order to provide orientation and training for outside living. (No refs.) - V. G. Votano.

2822 WILSON, GEORGE T. Status of recreation for the handicapped: School centered. In: American Association for Health, Physical Education and Recreation, & National Recreation and Park Association. *Physical Education and Recreation for Handicapped Children. Proceedings of a Study Conference on Research and Demonstration Needs*. Washington, D. C., 1969, p. 35-37.

School-centered recreation programs are usually conducted in day schools, special classes, or after school. Physical education activities should produce competencies which include mobility, manual skills, language, social relationships, and improved self image. Emphasis should be on the whole child, a sheltered environment, and the importance of affection. A primary task of research is to develop and perfect methods of evaluation with emphasis on measuring the enjoyment which the children receive from a program. Likes, dislikes, and attitudes must be measured, and people outside the program can be most effective in this type of evaluation. The most critical problems in establishing recreational programs for the handicapped are transportation and financing. (No refs.) V. G. Votano.

2823 FREEBERG, WILLIAM H. Research-Recreation camping for all handicapped. In: American Association for Health, Physical Education and Recreation & National Recreation and Park Association. *Physical Education and Recreation for Handicapped Children. Proceedings of a Study Conference on Research and Demonstration Needs*. Washington, D. C., 1969, p. 37-43.

Recreational camping offers a great deal in the education, rehabilitation, recreation, and social life of handicapped individuals. The experiences and values derived from

camping prior to 1960 were minimal because of a lack of research, facilities, staff, evaluation methods, and funds. One scientific research study concluded that camping resulted in enhanced self acceptance (measured by Hilden's Q-Instrument), few changes of interests, and increased social participation. Results from Kennedy Foundation studies indicated that parents favored camping, counselors gained more acceptance of the handicapped, and campers gained mentally, physically, and socially. The environment, selection of personnel, program content, and group living are important in the quality of the experience. Adequate means of testing and measurement are available for evaluation; however, there is a deficiency of qualified individuals to run such testing programs. More effort must be made to encourage students to enter recreation fields, and federal assistance could help establish a research center. (85-item bibliog.; 3 refs.) - V. G. Votano.

2824 BLATT, BURTON. Times' passage-unchanging times. In: American Association for Health, Physical Education and Recreation, & National Recreation and Park Association. *Physical Education and Recreation for Handicapped Children. Proceedings of a Study Conference on Research and Demonstration Needs*. Washington, D. C., 1969, p. 50-61.

Very little has been done to bring recreation and physical education to the mentally or physically retarded and research into these fields has been unproductive. Recommendations for research include the control of bias on the part of the researcher, the restriction of samples rather than variables in a study, the development of a better comprehension of the varieties and natures of physical education, and the appreciation of the importance of studies which involve individuals in the back wards of mental hospitals. Trained personnel in the medical and social areas should be conserved, and unskilled persons should be hired to assume routine responsibilities. The reservoirs of human talent must be utilized to alleviate our shortage of those people who deal with human welfare, and capable, competent workers must be employed without prejudice. (No refs.) - V. G. Votano.

2825 BRAATEN, JUNE, & LEE, ISABEL. *Swimming Program for the Trainable Retarded: Guide Number 1: Organisation and Administration of the Program*. Toronto, Canada, Canadian Association for the Mentally Retarded, 16 p.

An adequate swimming program can be of much value, physically and mentally, in the

growth and development of TMR and EMR children. In 1957, Canada conducted its first pilot-project study on swimming programs for the TMR which established guidelines, procedures, organization, and administration for an adequate swimming program to enhance the daily lives of their MR youth. The swimming program can either be a school function as part of the curriculum or one designated as an outside activity after school hours. The prime task of organizing such a program is to ascertain whether there is sufficient interest, to make the parents cognizant of the program and to obtain their approval for enrollment of their MR child. Sound organization is necessary by responsible persons interested in enriching experiences for the MR and many of these individuals will be professionals, volunteers, and members of community agencies and organizations. Swimming pool facilities and equipment must be taken into consideration and every phase of water safety carefully scrutinized. Staff, qualifications of personnel, committees responsible for transportation, insurance, cost, and public relations are important facets in maintaining a swimming program for the MR. Suggested forms and charts are available which indicate the division, direction, responsibility, and flow of authority. (No refs.) - S. Half.

2826 BRAATEN, JUNE, & LEE, ISABEL. *Swimming Program for the Trainable Retarded: Guide Number 2: Conducting the Program*. Toronto, Canada, Canadian Association for the Mentally Retarded, 33 p.

A detailed explanation of the program, staff, orientation, methods of instruction, evaluation, and a recording of progress of the swimming program for the TMR, and a record of the experience gained from the initial pilot project is presented. Volunteers and salaried staff can be utilized in the operation of a swimming program for MR children. Success depends upon the attitudes, approaches, and personalities of the instructors. Staff responsibilities vary, but should be well coordinated to achieve the purposes and goals set forth. Orientation of staff can be accomplished by observation, discussion groups, workshops, seminars, lectures, films, pamphlets, and familiarization with the facilities prior to commencement of the program. Outlined procedures should be carefully studied and followed while maintaining a comfortable degree of flexibility. Teaching suggestions are given; however, success depends

on the individual teacher's interest, patience, understanding and inventiveness. Specific instructions are given on how to best teach the MR various swimming strokes, positions, methods of breathing, arm and leg action, coordination, muscular control, body buoyancy, and deep water techniques and skills. Students should become familiar with proper use of water items and safety practices. Consistent and correct recording with an individual progress chart can serve as an indicator of the level of skill attained, the rate of progress, and the true ability of each pupil. Further analysis of the program can be ascertained by the design of master progress charts, percentage tables of skills exhibited, and statistical data to assess the students' skills as a group. (No refs.) S. Half.

2827 BRAATEN, JUNE, & LEE, ISABEL. *Swimming Program for the Trainable Retarded: Guide Number 3: Testing and Recognition*. Toronto, Canada, Canadian Association for the Mentally Retarded, 16 p.

This booklet is designed to help the swimming director and staff determine the exact direction that the swimming program for the TMR should follow. Testing and recognition of success are an integral phase of the program, and the Canadian Association for Retarded Children (CARC) has devised 2 types of awards for use in the swimming program. The report card, called Swimming Progress and Achievement Record, serves as a measure of the trainee's proficiency. The colorful Award Booklet is a tangible means for the MR to receive recognition of his success. A series of 7 booklets are available, one for each of the "learn to swim" advancements. The award system provides recognition of the child's success. Required materials for testing are available and can be obtained from the CARC or the Provincial Association. Additional work has to be done on testing and securing examiners, and this information will be forthcoming in a complete Testing Guide which will outline testing procedures for specific progressions. Standard Red Cross procedures can be used for testing, and the swimming director, at his discretion, may demonstrate each test item prior to the trainee performing. (No refs.) - S. Half.

FAMILY

2828 MURRAY, MAX A. (Mrs.) *Needs of Parents of Mentally Retarded Children*. New York, New York, National Association for Retarded Children, 1969, 16 p.

Major problem areas confronting families of MR children can be adequately met by honest, understanding, compassionate, and knowledgeable professionals. The lives of parents of MRs can be brightened by providing constructive information that will help them bring their children and themselves out of the shadow and give them the courage to carry their burden. Problems common to families of MR youngsters are the acceptance that they have a mentally deficient child, practical and intelligent use of available finances in terms of all family members, learning how to live successfully day-by-day with emotional traumatic experiences, the resolving of theological conflicts and guilt feelings within their own personal lives, and the ability to render sound decisions about the life-time care and supervision for their handicapped offspring. Parents frequently encounter professionals who lack needed knowledge concerning MR and who are emotionally immature and inexperienced. Inept advice given to an emotionally distraught parent can be worse than no help at all. Parents of MR children must be provided with constructive professional counseling throughout the child's life. (No refs.) - S. Half.

2829 SWINYARD, CHESTER A. The importance of family education and counseling. In: New York University Spina Bifida Group. *Comprehensive Care of the Child with Spina Bifida Manifesta*. Swinyard, Chester A., ed. New York, New York, New York University Medical Center (Rehabilitation Monograph 31), 1966, p. 114-130.

The parents of a child with spina bifida require the greatest psychological support when

the mother is recovering from the stress of childbirth and should receive initial education and counseling at that time. Initial counseling should be carried out by the patient's physician, while counseling on the specific details of a surgical procedure should be done by the specialist involved. Counseling should provide information on the nature and causes of spina bifida with myelomeningocele, the problems of the spina bifida child, early treatment, bowel and bladder management, and physical therapy. Genetic counseling should be based on the use of available risk figures and should present parents with an accurate picture of the problems of their child and the general principles of increased risk. Parent education should teach them how to acquaint siblings with the difficulties which will confront the spina bifida child. Parent group meetings can provide opportunities for parents to exchange experiences, share problems, and strengthen cooperative relationships between parents, professionals, and institutions. (No refs.) - J. K. Wyatt.

2830 WIGGLESWORTH, ROBERT. The conduct of the interview with the handicapped child and his family. In: Gardiner, Peter, MacKeith, Ronald, & Smith, Vernon, eds. *Aspects of Developmental & Paediatric Ophthalmology*. London, England, Spastics International Medical Publications, 1969, p. 106-109.

Situations which have to be dealt with concerning the blind or partially sighted include the sudden birth of a blind child, the suspicion of a possibility of blindness, the blindness diagnosed later in infancy, the development of blindness in later infancy, and the sudden blindness of a child due to accident or illness. In the case of blindness discovered at birth, the father should be told first, and he should be told of the disorder at the hospital, not by phone. The father must decide who is to report the disorder to the mother. Doctors, nurses, and

hospital personnel should help the parents adjust to their feelings. After the mother has been notified, she should be allowed to spend as much time as possible with the infant. (No refs.) - V. G. Votano.

2831 MILLER, LEE G. The seven stages in the life cycle of a family with a mentally retarded child. In: Washington. Institutions Department. *Proceedings of the Ninth Annual Research Meeting*. Olympia, Washington, Research Report, 2(2):78-81, 1969.

Stages in the life cycle of a family with an MR child are no MR children, diagnostic crisis and initial shock, prolonged dependency of the child, uncontrolled behavior of the child, adolescent development, academic failure and its implications for the future, and rejection by the community and lack of resources. The development of intra-family relationships is impeded in that the discovery of an MR child threatens the husband-wife union, distorts the mother-child relations, and disrupts the family balance by placing an excessive demand on the time, energy, affection, and attention of the mother. The father-child relationship does not develop, sibling relationships are skewed, and the social and personal lives of the parents are limited. The appearance of sexual and aggressive drives during adolescence elicits rejection from the community. The life cycle of a family with normal children moves toward reduced stress when the children leave home, while that of the family with an MR child moves toward increasing stress at each stage. Parents of MRs often seek help by requesting admission to an institution when stress loads threaten to exceed family strengths. The pattern of both the family and the institution is to wait for the total situation to deteriorate and then seek and provide help in an emergency situation. Steps which can intervene in this process of family breakdown and emergency include early casefinding, the provision of professional services for the total

family unit, a specific management program, short term placement as early as possible, and vocational training. (No refs.)

J. K. Wyatt.

2832 SMIRNOVA, A. N. *Vospitanie Umetvenno Otstalogo Rebenka Sem'e: Posobie dlya roditeli* (Education of Mentally Retarded Children in the Family: Handbook for Parents). Moscow, Union of Soviet Socialist Republics, Prosveshchenie Publishing House, 1967, 64 p., \$0.11.

Categories of MR, its pathological causes, and methods for its prevention are presented for parents of MR children. Many examples show the results of incorrect treatment and training of MR children and illustrate the secondary deficiencies that may occur. A large part of this book deals with overcoming the physical and mental retardation of Ss by educating them as to ideas, concepts, and feelings. In the appendices, the cooperation of the family and the school is discussed, and a plan is presented for observing the child's development by the use of daily routines. (No refs.) - R. K. Butler.

CONTENTS: MR Children; MR Children in the Family; Some Ways of Weakening and Overcoming Developmental Deficiencies in the MR Child; Education of the MR Child while Playing Games; Education of the MR Child during Work; How to Teach Moral Qualities to the MR Child; The Family and the School; Appendix 1 - Exemplary Plan of Observing the Development of the Child and the Direction of Work in the Family; Appendix 2 - Exemplary Daily Routine for Children of Early School Age; Appendix 3 - Exemplary Daily Routine for Children of Older School Age; Appendix 4 - Exemplary Lessons on Drawing, Modeling, and Handicrafts for MR Children of Pre-School and Early School Age.

PERSONNEL

- 2833 GARDNER, LYTT I. Genetic counseling. In: Gardner, Lytt I., ed. *Endocrine and Genetic Diseases of Childhood*. Philadelphia, W. B. Saunders, 1969, Chapter 22, p. 1023-1032.

Genetic counseling is both challenging and difficult, because the counselors must tamper with the patient's body-image and his concept of his own germ plasma as a continuum with his ancestors and descendants. Genetic counselors not only require clinical and medical knowledge of the highest order, but they also must be aware of the psychological aspects of the impact of a genetic disease on a family. Differential diagnosis between environmentally produced disease and genetic disease is the prerequisite to genetic counseling. Hereditary patterns of the genetic disorder involved and prognosis for the child should be discussed with parents so as to give a realistic picture of the problem. (19 refs.)
L. S. Ho.

- 2834 CLARK, D. F. A reassessment of the role of the clinical psychologist in the mental deficiency hospital. *Journal of Mental Subnormality*, 14(2):3-18, 1968.

In reassessing the role of the clinical psychologist in hospitals for MRs, the sociological characteristics of the hospital, the function of behavior testing, training and rehabilitation, therapy, research, and teaching are stressed. Sociological problems are partially caused by tightly scheduled programs involving the same groups of people for a longer period of time than is found in most other institutions. Patients and staff often have negative stereotype views of each other, thus creating a communication problem. The clinical psychologist must utilize concepts of social psychology in developing teamwork for the purpose of reaching common goals. Staff meetings, and patient groupings for therapy, discussion, or simple instructions help to achieve teamwork. Because of the interrelatedness of social and cognitive factors, the clinician must use appropriate tests of emotionality, impulsiveness, and modes of social and personal adjustments in

addition to IQ scores before prescribing placement and treatment. Considerable time must be spent in social and job training with community living as the goal for some patients. Teaching machines, programmed instruction, and operant conditioning techniques are helpful. Research, especially in the area of psychophysiological techniques, should be pursued. Teaching the staff about psychology and its clinical application must be deliberately planned and appropriately executed according to its intentions. (53 refs.)

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- 2835 ANDERSON, ROBERT M., & LITTLE, HARRY A. A practicum oriented teacher education program. *Education and Training of the Mentally Retarded*, 3(2):75-79, 1968.

An undergraduate university program for future teachers of MR provides practicum experience from the freshman year through student teaching and answers a frequent criticism that MR education students do not have enough contact with the type of pupils they will teach. A campus laboratory school has special education classes for EMRs and TMRs from kindergarten through high school. University freshmen observe in local and laboratory schools, watch films of actual class techniques, and work in a summer camp for MRs. Sophomores have practical classes coupled with observation and field work in local clinics, institutions, and agencies. A valuable new sophomore field experience includes living a week in the state MR school; a tour of facilities, observation of staff patterns, study of selected residents' records, observation of the participation of various staff members in a case. Juniors participate in MR classes, keep records, and learn curriculum trends, methods, teaching materials, and curriculum overviews. Seniors teach for 9 weeks in area schools for a competency in TMR or EMR and 18 weeks for both. (3 refs.)
M. Pleasinger.

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Normal, Illinois 61761

2836 HAMERLYNCK, LEO A., MARTIN, JACK W., & ROLLAND, JOHN C. Systematic observation of behavior: A primary teacher skill. *Education and Training of the Mentally Retarded*, 3(1):39-42, 1968.

Teachers should be trained to observe MR classroom students for task-oriented behavior or lack of it. In a summer secondary level session for 17 MRs, 4 were chosen for observation, each by one team of 4 to 5 graduate teachers of the MR. They observed for 1 1/2 hours for three 5-day weeks. Although the class teacher had stated that no child had problem behavior, the 4 Ss spent an average

of 45% of their time in task-oriented behavior--listening to the teacher, reading the problem, and writing a product. The non-task oriented behavior was not disruptive to the class--looking around, putting head on desk--and so elicited no teacher response. Effective teaching was impossible for the Ss, and task observation had not been stressed in teacher training. Teachers need to develop observation skills which will aid in reinforcement of task-oriented behavior. (7 refs.)

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PUBLICATIONS SCANNED

The following publications are scanned regularly for articles pertinent to mental retardation.

AAUP Bulletin
 ACLD Items of Interest (Association for Children
 with Learning Disabilities)
 ACT (American College Testing Program) Research
 Reports
 AIA (Architectural Institute of America)
 ALA Bulletin (American Library Association)
 AMS Bulletin (American Montessori Society)
 ASHA: A Journal of the American Speech and Hearing
 Association
 ATA Magazine
 ATA News (Alberta Teachers Association)
 AV (Audio-Visual) Communication Review
 About Education
 Abstracts for Social Workers
 Academic Therapy Quarterly
 Acta Anatomica
 Acta Biologicae Experimentalis
 Acta Chirurgiae Plasticae
 Acta Endocrinologica
 Acta Geneticae Medicae et Gemellologiae
 Acta Medica Belgica
 Acta Medica Scandinavica
 Acta Morphologica
 Acta Morphologica Academiae Scientiarum Hungaricae
 Acta Neurochirurgica
 Acta Neurologica et Psychiatrica Belgica
 Acta Neurologica Scandinavica
 Acta Neuropathologica
 Acta Obstetrica et Gynaecologia Scandinavica
 Acta Ophthalmologica
 Acta Oto-Laryngologica
 Acta Paediatrica Belgica
 Acta Paediatrica Academiae Scientiarum Hungaricae
 Acta Paediatrica Scandinavica
 Acta Paedopsychiatrica
 Acta Pathologica et Microbiologica Scandinavica
 Acta Physiologica Latino Americana
 Acta Physiologica Polonica
 Acta Psiquiatrica y Psicologica de American
 Latina
 Acta Psychiatrica Scandinavica
 Acta Psychologica, Amsterdam
 Acta Sociologica
 Activitas Nervosa Superior
 Administrative Science Quarterly
 Administrator's Notebook
 Adolescence
 Adult Education
 Adult Education News
 Adult Leadership
 Aerospace Medicine
 Aerzliche Forschung
 Agricultural Education Magazine
 Alabama School Journal
 Alabama State Teachers Association Journal
 Alaska Teacher
 Alberta Journal of Educational Research
 Alberta Psychologist
 Alma Mater
 Alpha Delta Kappa
 Amentia
 American Latina, Brazil
 American Annals of the Deaf
 American Anthropologist
 American Association for Health, Physical Education,
 and Recreation Research Quarterly
 American Association of Colleges for Teacher
 Education Yearbook

American Association of School administrators
 Official Report
 American Behavioral Scientist
 American Biology Teacher
 American Child
 American Corrective Therapy Journal
 American Council on Industrial Arts Teacher
 Education Yearbook
 American Ecclesiastical Review
 American Education
 American Educational Research Journal
 American Foundation for the Blind, Research
 Bulletin
 American Heart Journal
 American Institute of Architects Journal
 American Journal of Art Therapy
 American Journal of Cardiology
 American Journal of Clinical Hypnosis
 American Journal of Clinical Nutrition
 American Journal of Clinical Pathology
 American Journal of Correction
 American Journal of Digestive Diseases
 American Journal of Diseases of Children
 American Journal of Economics and Sociology
 American Journal of Epidemiology
 American Journal of Hospital Pharmacy
 American Journal of Human Genetics
 American Journal of Medical Sciences
 American Journal of Medicine
 American Journal of Mental Deficiency
 American Journal of Nursing
 American Journal of Obstetrics and Gynecology
 American Journal of Occupational Therapy
 American Journal of Ophthalmology
 American Journal of Optometry and Archives of
 American Academy of Optometry
 American Journal of Orthodontics
 American Journal of Orthopsychiatry
 American Journal of Pathology
 American Journal of Physical Anthropology
 American Journal of Physical Medicine
 American Journal of Physiology
 American Journal of Proctology
 American Journal of Psychiatry
 American Journal of Psychoanalysis
 American Journal of Psychology
 American Journal of Psychotherapy
 American Journal of Public Health
 American Journal of Roentgenology
 American Journal of Roentgenology, Radium Therapy,
 and Nuclear Medicine
 American Journal of Sociology
 American Journal of Surgery
 American Journal of Tropical Medicine and Hygiene
 American Music Teacher
 American Psychologist
 American Quarterly
 American Review of Respiratory Diseases
 American School and University
 American School Board Journal
 American School News
 American Sociological Review
 American Sociologist
 American Surgeon
 American Teacher
 American Teacher Magazine
 American Vocational Journal
 Anaesthesia
 Analyse et Prevision, France

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Anatomical Record
 Andover Newton Quarterly
 Anesthesia and Analgesia: Current Researches
 Anesthesia Progress
 Anesthesiology
 Angiology
 Anglican Theological Review
 Animal Behavior
 Annales d'Endocrinologie
 Annales de Genetique
 Annales de Pediatrie
 Annales Medico-Psychologiques
 Annales Paediatricae Fenniae
 Annali Sociologia, Italy
 Annals Institute de Pasteur, Paris
 Annals of Allergy
 Annals of Human Genetics
 Annals of Internal Medicine
 Annals of Rheumatic Diseases
 Annals of Surgery
 Annals of the New York Academy of Sciences
 Annals of Thoracic Surgery
 Annee Psychologique
 Annual of Animal Psychology, Tokyo
 Annual Review of Psychology
 Antibiotiki
 Antioch Review
 Aportes
 Applied Social Studies
 Architects' Exchange
 Architectural Forum
 Architectural Record
 Architectural Review
 Archives Francaises de Pediatrie
 Archives Italiennes de Biologie
 Archives of Biochemistry and Biophysics
 Archives of Dermatology
 Archives of Disease in Childhood
 Archives of Environmental Health
 Archives of General Psychiatry
 Archives of Internal Medicine
 Archives of Neurology
 Archives of Ophthalmology
 Archives of Otolaryngology
 Archives of Pathology
 Archives of Physical Medicine
 Archives of Surgery
 Archiv fur die gesamte Psychologie
 Archiv fur Geschwulstforschung
 Archiv fur Kinderheilkunde
 Archiv fur Klinische und experimentelle Ohren-
 Nasen-und Kehlkopfheilkunde
 Archiv fur Psychiatrie und Nervenkrankheiten
 Archivio di Psicologia, Neurologia e Psichiatria
 Archivos de Crimonologia, Neuropsiquiatria y
 Disciplinas Conexas
 Arhiv za Higijenu Radu i Toksikologiju
 Arithmetic Teacher
 Arizona Teacher
 Arquivos de Neuro-Psiquiatria
 Art Education
 Arthritis and Rheumatism
 Art Quarterly
 Arts and Activities
 Association for Student Teaching Yearbook
 Association for Supervision and Curriculum
 Development Yearbook
 Athletic Journal
 Audiovisual Instruction
 Audio-Visual Language Journal
 Audio-Visual Media
 Australasian Annals of Medicine
 Australian Children Limited
 Australian Journal of Adult Education
 Australian Journal of Education
 Australian Journal of Experimental Biology
 and Medical Science
 Australian Journal of Psychology
 Australian Journal of Social Issues
 Australian Outlook
 Australian Paediatric Journal

BINOP: Bulletin de l'Institut National d'Etude
 du Travail et d'Orientation Professionnelle
 Balance Sheet
 Behavior

Behavioral Science
 Behavior Research Methods and Instrumentation
 Behaviour Research and Therapy
 Bibliotheca Psychiatrica et Neurologica
 Bien-Etre Social Canadien
 Bildung und Erziehung
 Biochemical and Biophysical Research communications
 Biochemical Journal
 Biochemical Medicine
 Biochemistry
 Biochimica et Biophysica Acta, Amsterdam
 Biofizika
 Biologia Neonatorum
 Biological Psychiatry
 Biometrics
 Birth Defects Original Article Series
 Blood
 Blut
 Boletin Informativo (Instituto Nacional de
 Psiquiatria Infantil)
 Boletin Informativo del Instituto Neurologico
 de Guatemala
 Bollettino di Psicologia Applicata
 Brain
 Brain, Behavior and Evolution
 Brain Research
 British Heart Journal
 British Journal for the Philosophy of Science
 British Journal of Clinical Practice
 British Journal of Criminology
 British Journal of Dermatology
 British Journal of Educational Psychology
 British Journal of Educational Studies
 British Journal of Industrial Medicine
 British Journal of Mathematical and Statistical
 Psychology
 British Journal of Medical Psychology
 British Journal of Ophthalmology
 British Journal of Pharmacology
 British Journal of Preventive and Social Medicine
 British Journal of Psychiatric Social Work
 British Journal of Psychiatry
 British Journal of Psychology
 British Journal of Radiology
 British Journal of Social and Clinical Psychology
 British Journal of Sociology
 British Journal of Surgery
 British Medical Journal
 Broadcaster (Newsletter of the Beatrice State
 home, Beatrice, Nebraska)
 Bulgarska Akademiya Na Naukite
 Bulletin (Council of Social and Psychological
 Research, Calcutta)
 Bulletin de l'Association Internationale de
 Psychologie Appliquee
 Bulletin de Psychologie Scolaire et de l'Orientation
 Bulletin des Ecoles Primaires
 Bulletin du C.E.R.P.
 Bulletin of Suicidology
 Bulletin of the Atomic Scientists
 Bulletin of the British Psychological Society
 Bulletin of the Dental Guidance Council for
 Cerebral Palsy
 Bulletin of the Department of Education (Mass.)
 Bulletin of the Institute of Child Study
 Bulletin of the International Bureau of Education
 Bulletin of the Los Angeles Neurological Society
 Bulletin of the Menninger Clinic
 Bulletin of the National Association of Secondary
 School Principals
 Bulletin of the New York Academy of Medicine
 Bulletin of the School of Education (Indiana U.)
 Bulletin of Tokyo Dental College
 Business Education Forum
 Business Education World
 Byulletin' Eksperimental' noi Biologii i Meditsiny

CTA (California Teachers Association) Journal
 Cahiers de Psychologie
 Cahiers de Sociologie Economique
 California Education
 California Elementary School Administrators
 Association Monographs
 California Journal of Educational Research
 California Mental Health Research Digest
 California Medicine

PUBLICATIONS SCANNED

California School Boards
California School Employee
California School Libraries
Canada's Mental Health
Canada's Mental Health Supplement
Canadian Administrator
Canadian Anaesthetists' Society Journal
Canadian Audio Visual Review
Canadian Education and Research Digest
Canadian Journal of Behavioral Science
Canadian Journal of Biochemistry
Canadian Journal of Corrections
Canadian Journal of Physiology and Pharmacology
Canadian Journal of Psychology
Canadian Journal of Surgery
Canadian Journal of Theology
Canadian Medical Association Journal
Canadian Nurse
Canadian Psychiatric Association Journal
Canadian Psychologist
Canadian Review of Sociology and Anthropology
Canadian School Journal
Canadian Welfare
Cancer
Cancer Research
Cardiovascular Research
Case Conference
Casopsis Lekaru Ceskych
Catholic Charities Review
Catholic Educational Review
Catholic Educator
Catholic High School Quarterly
Catholic Psychological Record
Catholic School Journal
Central Missouri Synthesis in Mental Retardation
Centro Ricerche Biopsichiche
Cerebral Palsy Journal
Ceskoslovenska Psychiatrie
Ceskoslovenska Psychologie
Challenge
Changing Education
Character Potential
Cheshire Smile
Child and Family
Child Development
Child Development Abstracts and Bibliography
Child Education
Child Health in Israel
Childhood Education
Children
Children Limited
Children's House
Child Study
Child Study Center Bulletin (State University
Coll. New York, Buffalo)
Child Welfare
Chile Instituto Nacional Boletín
Chirurg
Christianity and Crisis
Christianity Today
Christian Scholar
Church Teacher
Circulation
Circulation Research
Claremont Reading Conference Yearbook
Classical Journal
Clearing House
Clearing House Journal
Clergy Review
Clinica Chimica Acta
Clinical and Experimental Immunology
Clinical Chemistry
Clinical Pediatrics
Clinical Pharmacology and Therapeutics
Clinical Proceedings of Children's Hospital of the
District of Columbia
Clover Leaves-Observer
College and University
College English
College of Education Record
College Student Survey
Colorado Education Review
Color Engineering
Commentary
Community Health
Community Mental Health Journal
Comparative Education (Oxford)

Comparative Education Review
Comprehensive Psychiatry
Concordia Historical Institute Quarterly
Conditional Reflex
Conference Board Record
Conference on Reading (University of Chicago)
Confinia Neurologica
Confinia Psychiatrica
Connecticut Health Bulletin
Connecticut Teacher
Contemporary Education
Contemporary Psychoanalysis
Contemporary Psychology
Contributi dell'Istituto di Psicologia
Cornell Journal of Social Relations
Corrective Psychiatry and Journal of Social
Therapy
Cortex
Council for Research in Music Education
Counselor Education and Supervision
Courier
Crime and Delinquency
Culture and Education
Current Anthropology
Current Contents: Life Sciences
Current Therapeutic Research
Curriculum and Materials
Curriculum Bulletin
Curriculum Leadership
Curriculum Report
Cybernetics
Cytochemistry
DSH Abstracts
Daedalus
Dapim Refuim
Defence Science Journal
Deficiency Mentale/Mental Retardation
Delaware Association for Retarded Children—News
Delaware School Journal
Der Landarzt
Deutsche Medizinische Wochenschrift
Deutsches Aezzteblatt
Deutsche Zeitschrift für Nervenheilkunde
Developmental Medicine and Child Neurology
Developmental Psychology
Diabetes
Diabetologia
Diagnostica: Zeitschrift für Psychologische
Diagnostik
Didaktometrie
Die Medizinische
Die Rehabilitation
Difesa Sociale
Digest of the Mentally Retarded
Diseases of the Chest
Diseases of the Colon and Rectum
Diseases of the Nervous System
Dissertation Abstracts
Doklady Akademii Nauk SSSR
Doshkoi' noe Vospitanie
ERIC Research in Education
ETC: A Review of General Semantics
ETV Reporter
Economic Development and Cultural Change
Education
Education (London)
Education (USA)
Education Age
Educational Administration Quarterly
Educational and Psychological Interactions
Educational and Psychological Measurement
Educational Bulletin (Iowa)
Educational Development
Educational Forum
Educational Horizons
Educational Leadership
Educational Magazine
Educational Perspectives
Educational Psychologist
Educational Psychology
Educational Record
Educational Record of Province of Quebec
Educational Records Bureau Bulletins

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Educational Research (British)
Educational Researcher
Educational Review
Educational Sciences
Educational Screen AV (Audio-Visual) Guide
Educational Technology
Educational Television
Educational Theatre Journal
Educational Theory
Educational Times
Education and Culture
Education and Psychology Review
Education and Training of the Mentally Retarded
Education Canada
Education Digest
Education Index
Education in France
Education Panorama
Education Quarterly
Education Review
Education Today
Educatio Italiano
Educators Guide to Media and Methods
Ek'sperimental'nyy Klinicheskiy Zhurnal
Electroencephalography and Clinical Neurophysiology
Elementary English
Elementary School Guidance and Counseling
Elementary School Journal
Employment Service Review
Encephale
Encounter
Endocrinology
Enfance
English in Education
English Journal
English Language Teaching
Environment
Environmental Research
Epilepsia
Ergonomics
ERIC-IRCD Bulletin
Erziehung und Unterricht
European Neurology
European Surgical Research
Evangelische Theologie, Germany
Evolution Psychiatrique
Exceptional Child Education Abstracts
Exceptional Children
Excerpta Criminologica
Experientia
Experimental Gerontology
Experimental Neurology
Explorations in Entrepreneurial History
Expository Times
Eye, Ear, Nose and Throat Monthly

Family Care Newsletter
Family Law Quarterly
Family Life Coordinator
Family Process
Farmakologiya o Tpisloip'pgoua
Federation Proceedings
Fertility and Sterility
Film Quarterly
Fiziologicheskii Zhurnal SSSR
Flight Safety
Florida Education
Focus on Exceptional Children
Folia Psychiatrica et Neurologica Japonica
Forecast for Home Economics
Fortschritte auf dem Gebiete der Rotgenstrahlen
un der Nuklearmedizin
Forum
Forum for the Discussion of New Trends in
Education
Forward Trends
Foundations
France Medicale
Free University Quarterly, Holland
Franch Review

GAP (Group for the Advancement of Psychiatry)
Report
Gastroenterology
Gastrointestinal Endoscopy

Gawein
Geburtshilfe und Frauenheilkunde
General Practice
Genetic Psychology Monographs
Genetika
Georgia Educational Journal
Geriatrics
German Medical Monthly
German Quarterly
Gerontologia
Gerontologia Clinica
Gerontologist
Gifted Child Quarterly
Gordon Review
Grade Teacher
Graduate Research in Education and Related
Disciplines
Group Psychotherapy
Guidance Journal
Gynaecologia
Gynakologie

Hachinuch
Harefuah
Harpers Magazine
Harvard Educational Review
Harvard Theological Review
Headache
Head Teachers Review
Health, Education and Welfare Indicators
Health Education Journal
Health Laboratory Science
Heilpädagogische Forschung
Heilpädagogische Werkblätter
Helping Services Dorum
Helvetica Paediatrica Acta
Hereditas
Hibbert Journal
High Points
High School Journal
Hispania
Hispanic Review
History of Education Quarterly
History of Religions
Hjertebladet
Homiletic and Pastoral Review
Hommes et Techniques
Hospital and Community Psychiatry
Hospital Management
Hospital Practice
Hospital Progress
Hospita (Rio de Janeiro)
Hospitals
Human Biology: An International Record of Research
Human Development
Human Factors
Humangenetik
Human Heredity
Human Organization
Human Potential
Human Relations
Humanitas (Brescia)
HumRRO Professional Paper
HumRRO Technical Report
Hygiene Mentale

IAPPW Journal—International Association of
Pupil Personnel Workers
ICRH (Information Center, Recreation for the
Handicapped) Newsletter
IEEE Transactions on Education
IEEE Transactions on Human Factors in Electronics
IMRD (Institute on Mental Retardation and Intellectual
Development) Papers and Reports
Illinois Education
Illinois Journal of Education
Illinois Medical Journal
Illinois School Research
Illinois Schools Journal
Illinois State University Journal
Immunology
Impact of Science on Society
Improving College and University Teaching
Independent School Bulletin
Indiana Teacher

PUBLICATIONS SCANNED

Indiana University School of Education Bulletin
 Indian Education
 Indian Educational Review
 Indian Journal of Educational Administration and Research
 Indian Journal of Extension Education
 Indian Journal of Mental Retardation
 Indian Journal of Psychology
 Indian Journal of Social Work
 Indian Journal of Theology
 Indian Psychological Review
 Individual Psychologist
 Industrial Arts and Vocational Education/Technical Education
 Industrial Relations
 Information Psychologique
 Inquiry
 Insight: Quarterly Review of Religion and Mental Health
 Institute for Educational Innovation Bulletin
 Institute of Dream Research Monograph Series
 Instructor
 Instructor Magazine
 Instrumentalist
 Integrated Education
 International Archives of Allergy and Applied Immunology
 International Bureau of Education Bulletin
 International Child Welfare Review
 Internationales Archiv für Gewerbepathologie und Gewerbehygiene
 International Journal for the Education of the Blind
 International Journal of Clinical and Experimental Hypnosis
 International Journal of Fertility
 International Journal of Group Psychotherapy
 International Journal of Neuropsychology
 International Journal of Neuropsychiatry
 International Journal of Nursing Studies
 International Journal of Offender Therapy
 International Journal of Parapsychology
 International Journal of Psychiatry
 International Journal of Psycho-Analysis
 International Journal of Psychology
 International Journal of Radiation Biology
 International Journal of Social Psychiatry
 International Journal of Sociometry and Sociatry
 International Nursing Review
 International Pharmacopsychiatry
 International Rehabilitation Review
 International Review of Community Development
 International Review of Education
 International Review of Missions
 International Social Sciences Journal
 International Social Security Review
 International Social Work
 International Yearbook of Education
 Irish Journal of Medical Science
 Israel Annals of Psychiatry and Related Disciplines
 Israel Annals of Psychology and Related Disciplines
 Israel Journal of Medical Sciences
 Italian Studies
 Italica

Japanese Journal of Child Psychiatry
 Japanese Journal of Educational Psychology
 Japanese Journal of Experimental Medicine
 Japanese Journal of Human Genetics
 Japanese Journal of Medical Science and Biology
 Japanese Journal of Pharmacology
 Japanese Psychological Research
 Jewish Education
 Jewish Parent
 Jewish Social Studies
 Johns Hopkins Medical Journal
 Journal de Genetique Humaine
 Journal de Physiologie
 Journal de Psychologie Normale et Pathologique
 Journal for Social Research
 Journal for Special Educators of The Mentally Retarded
 Journal for the Scientific Study of Religion
 Journalism Quarterly

Journal of Abnormal Psychology
 Journal of Aesthetic Education
 Journal of Aesthetics and Art Criticism
 Journal of Air Pollution Control Association
 Journal of Allergy
 Journal of American Indian Education
 Journal of Analytical Psychology
 Journal of Applied Behavioral Science
 Journal of Applied Behavior Analysis
 Journal of Applied Physiology
 Journal of Applied Psychology
 Journal of Architectural Education
 Journal of Arkansas Education
 Journal of Association for the Study of Perception
 Journal of Asthma Research
 Journal of Auditory Research
 Journal of Bacteriology
 Journal of Biological Psychology
 Journal of Biosocial Science
 Journal of Bone and Joint Surgery
 Journal of Business Education
 Journal of Cell Biology
 Journal of Chemical Education
 Journal of Child Psychology and Psychiatry and Allied Disciplines
 Journal of Chronic Diseases
 Journal of Church and State
 Journal of Clinical Endocrinology
 Journal of Clinical Endocrinology and Metabolism
 Journal of Clinical Investigation
 Journal of Clinical Pathology
 Journal of Clinical Pharmacology
 Journal of Clinical Psychology
 Journal of College Placement
 Journal of Communication
 Journal of Communication Disorders
 Journal of Comparative and Physiological Psychology
 Journal of Comparative Neurology
 Journal of Conflict Resolution
 Journal of Consulting and Clinical Psychology
 Journal of Contemporary Psychotherapy
 Journal of Correctional Education
 Journal of Counseling Psychology
 Journal of Creative Behavior
 Journal of Criminal Law, Criminology and Police Science
 Journal of Dentistry for Children
 Journal of Ecclesiastical History
 Journal of Education (Boston)
 Journal of Education (Halifax)
 Journal of Educational Administration
 Journal of Educational Data Processing
 Journal of Educational Measurement
 Journal of Educational Psychology
 Journal of Educational Research
 Journal of Education for Social Work
 Journal of Emotional Education
 Journal of Employment Counseling
 Journal of Endocrinology
 Journal of Engineering Psychology
 Journal of Environmental Health
 Journal of Existentialism
 Journal of Experimental Child Psychology
 Journal of Experimental Education
 Journal of Experimental Medicine
 Journal of Experimental Psychology
 Journal of Experimental Research in Personality
 Journal of Experimental Social Psychology
 Journal of Family Welfare
 Journal of General Education
 Journal of General Psychology
 Journal of Genetic Psychology
 Journal of Geography
 Journal of Gerontology
 Journal of Health and Social Behavior
 Journal of Health, Physical Education, and Recreation
 Journal of Heredity
 Journal of Higher Education
 Journal of Home Economics
 Journal of Human Relations
 Journal of Human Resources
 Journal of Immunology
 Journal of Individual Psychology
 Journal of Industrial Arts Education
 Journal of Investigative Dermatology
 Journal of Jewish Communal Service

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Journal of Laboratory and Clinical Medicine
 Journal of Laryngology and Otolaryngology
 Journal of Learning Disabilities
 Journal of Lipid Research
 Journal of Marriage and the Family
 Journal of Mathematical Psychology
 Journal of Medical Education
 Journal of Medical Genetics
 Journal of Medicinal Chemistry
 Journal of Mental Deficiency Research
 Journal of Mental Subnormality
 Journal of Motor Behavior
 Journal of Music Therapy
 Journal of Negro Education
 Journal of Nervous and Mental Disease
 Journal of Neurochemistry
 Journal of Neurology, Neurosurgery and Psychiatry
 Journal of Neuropathology and Experimental
 Neurology
 Journal of Neurophysiology
 Journal of Neurosurgery
 Journal of Neuro-Visceral Relations
 Journal of Nuclear Medicine
 Journal of Nutrition
 Journal of Nutrition Education
 Journal of Obstetrics and Gynecology of the
 British Commonwealth
 Journal of Occupational Medicine
 Journal of Oral Surgery
 Journal of Oral Surgery, Oral Medicine and Oral
 Pathology
 Journal of Parapsychology
 Journal of Pastoral Care
 Journal of Pathology and Bacteriology
 Journal of Pediatrics
 Journal of Periodontal Research
 Journal of Personality
 Journal of Personality and Social Psychology
 Journal of Pharmaceutical Sciences
 Journal of Pharmacology and Experimental
 Therapeutics
 Journal of Physical Education
 Journal of Programmed Instruction
 Journal of Projective Techniques and Personality
 Assessment
 Journal of Psychiatric Nursing and Mental Health
 Services
 Journal of Psychiatric Research
 Journal of Psychological Researches
 Journal of Psychology
 Journal of Psychopharmacology
 Journal of Psychosomatic Research
 Journal of Public Health Dentistry
 Journal of Reading
 Journal of Rehabilitation
 Journal of Rehabilitation in Asia
 Journal of Religion and Health
 Journal of Research and Development in Education
 Journal of Research in Music Education
 Journal of School Health
 Journal of School Psychology
 Journal of Secondary Education
 Journal of Social Issues
 Journal of Social Psychology
 Journal of Social Work Process
 Journal of Special Education
 Journal of Speech and Hearing Disorders
 Journal of Speech and Hearing Research
 Journal of State School Systems Development
 Journal of Surgical Research
 Journal of Teacher Education
 Journal of the Academy of Psychologists in
 Marital Counseling
 Journal of the Acoustical Society of America
 Journal of the American Academy of Child
 Psychiatry
 Journal of American Dental Association
 Journal of the American Dietetic Association
 Journal of the American Geriatrics Society
 Journal of the American Institute of Hypnosis
 Journal of the American Medical Association
 Journal of the American Optometric Association
 Journal of the American Physical Therapy
 Association
 Journal of the American Psychoanalytic
 Association
 Journal of the American Society for Physical
 Research

Journal of the American Society of Psychosomatic
 Dentistry and Medicine
 Journal of the American Statistical Association
 Journal of the College of General Practice
 Journal of the Experimental Analysis of Behavior
 Journal of the Hillside Hospital
 Journal of the History of the Behavioral Sciences
 Journal of the Irish Medical Association
 Journal of the National Cancer Institute
 Journal of the National Medical Association
 Journal of the Neurological Sciences
 Journal of the New York State School Boards
 Association
 Journal of the Optical Society of America
 Journal of the Reading Specialist
 Journal of the Scottish Society for Mentally
 Handicapped Children
 Journal of the Society for Psychical Research
 Journal of the Wisconsin State Dental Society
 Journal of Thoracic and Cardiovascular Surgery
 Journal of Thought
 Journal of Trauma
 Journal of Tropical Pediatrics
 Journal of Typographic Research
 Journal of Urology
 Journal of Verbal Learning and Verbal Behavior
 Journal of Virology
 Journal of Vocational and Educational Guidance
 Judaism
 Junior College Journal
 Jyväskylä Studies in Education, Psychology and
 Social Research

Kansas Schools
 Kansas Studies in Education (Kansas U.)
 Kansas Teacher
 Kappa Delta Pi Record
 Kasvatusopillinen Aikakauskija
 Kentucky Education News
 Kentucky School Journal
 Kenya Education Journal
 Khirurgia, Moscow
 Kleine Fachbuchreihe (Kuratorium für
 Verkehrssicherheit, Vienna)
 Klinische Medizin, Vienna
 Klinicheskaya Meditsina
 Klinische Wochenschrift, Berlin
 Kölner Zeitschrift für Soziologie und
 Sozialpsychologie
 Kulturno-Prosvetitel'naya Rabota
 Kwartalnik Pedagogiczny
 Kyklos

LTSH Observer (Lynchburg Training School and
 Hospital)
 Laboratory Investigation
 Lakartidningen, Stockholm
 Lancet
 Language and Speech
 Language Learning
 Larartidningen
 Laryngoscope
 Laval Medical, Quebec
 Lebenshilfe
 Liberal Education
 Library Journal
 Library Journal and School Library Journal
 Library Quarterly
 Life Sciences
 Linguistics
 London Quarterly and Holborn Review
 Louisiana Schools
 Lumen Vitae, Belgium
 Lupta de Clasa, Rumania
 Lutheran Quarterly
 Lutheran World

Magyar Pszichologiai Szemle
 Main Currents in Modern Thought
 Malaysian Journal of Education
 Manas
 Manitoba Journal of Educational Research
 Maryland Teacher
 Massachusetts Teacher
 Mathematics Teacher

PUBLICATIONS SCANNED

Mayo Clinic Proceedings
 McGill Journal of Education
 Measurement and Evaluation in Guidance
 Medicine Infantile
 Medical and Biological Illustration
 Medical Care
 Medical Journal
 Medical Journal of Australia
 Medical Research Engineering
 Medical Social Work (London)
 Medical Thoracalis
 Medical World News
 Medicine
 Medizinische Klinik, Munich
 Medizinische Welt, Stuttgart
 Megamot
 Menninger Quarterly
 Mennonite Quarterly Review
 Mensagem da APAE
 Mensch und Arbeit
 Mens en Onderneming
 Mental Health (National Association for
 Mental Health, London)
 Mental Health Digest
 Mental Hygiene
 Mental Retardation Abstracts
 Mental Retardation in Illinois
 Mental Retardation/MR
 Mental Retardation News
 Merrill-Palmer Quarterly
 Metabolism
 Metabolism, Clinical and Experimental
 Michigan Education Journal
 Michigan Journal of Secondary Education
 Michigan Quarterly Review
 Microbios
 Middle States Association of Colleges and
 Secondary Schools Proceedings
 Milbank Memorial Fund Quarterly
 Military Medicine
 Mind Over Matter
 Minerva
 Minerva Medica
 Minerva Medical Journal
 Minerva Pediatrica, Turin
 Minnesota Journal of Education
 Minnesota Student Journal
 Minnesota Studies in Vocational
 Rehabilitation
 Mississippi Educational Advance
 Mississippi Educational Journal
 Missouri Journal of Research in Music
 Education
 Modern Hospital
 Modern Language Journal
 Modern Vocational Trends
 Monographies Francaises de Psychologie
 Monographs of the Society for Research in
 Child Development
 Montana Education
 Monthly Labor Review
 Motive
 Multivariate Behavioral Research
 Multivariate Behavioral Research Monographs
 Munchener Medizinische Wochenschrift, Munich
 Music Educators Journal
 Music Journal
 Muslim World
 Muzika, Rumania

NCEA (National Catholic Educational
 Association) Bulletin
 NEA (National Educational Association)
 Research Bulletin
 NEA (National Education Association)
 Research Report
 NEA (National Education Association) Journal
 NIH (National Institutes of Health) Record
 Nachal'naya Shkola
 National Association of Secondary School
 Principals Bulletin
 National Association of Student Councils
 Yearbook
 National Association of Women Deans and
 Counselors Journal
 National Business Education Quarterly

National Business Education Yearbook
 National Council for the Social Studies
 National Council of Teachers of Mathematics
 Yearbook
 National Educational TV
 National Education Association Addresses and
 Proceedings
 National Elementary Principal
 National Institute of Industrial Psychology
 Paper
 National Merit Scholarship Corporation
 Research Reports
 National School Law Reporter
 National Society for the Study of Education
 Yearbook
 Nation's Schools
 Nature
 Nauka i Religiya
 Nauka i Zhizn'
 Nebraska Symposium on Motivation
 Nederlands Tijdschrift voor de Psychologie
 en haar Grensgebieden
 Nederlands Tijdschrift voor Geneeskunde
 Negro Educational Review
 Nervenarzt
 Neue Zeitschrift fur Systematische Theologie
 Neurologia, Psihiatria, Neurochirurgia
 Neurology
 Neuropsychiatria
 Neuropsychiatria Infantile
 New Education (London)
 New England Association Review
 New England Journal of Medicine
 New Outlook for the Blind
 New Research in Education
 New Scholasticism
 Newsletter of Chaplains and Other Religious
 Workers Subsection AAMD
 Newsletter of the International Union for
 Child Welfare
 Newsletter of the New Jersey Association for
 Brain Injured Children
 Newsletter of the Tennessee Association for
 Retarded Children and Adults
 Newsletter-The Aid for Retarded Children,
 Inc. of Stamford, Connecticut
 New York City Board of Education Curriculum
 Bulletins
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 New York Society for the Experimental Study
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 New York State Journal of Medicine
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 North Carolina ARC News
 North Carolina Education
 North Central Association Quarterly
 North Dakota Teacher
 Northeastern Studies in Vocational
 Rehabilitation
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 Nouvelle Revue Theologique
 Nova et Vetera, France
 Nursing Mirror
 Nursing Outlook
 Nursing Research
 Nutrition Reviews

Obstetrics and Gynecology
 Occupational Mental Health Notes
 Occupational Outlook Quarterly
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 Ohio State Medical Journal
 Oklahoma Teacher
 Ontario Journal of Educational Research
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 Onze Taak
 Operations Research
 Ophthalmologica

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Oral Surgery, Oral Medicine and Oral Pathology
Oregon Education
Oregon Health Bulletin
Organizational Behavior and Human Performance
Orientamenti Pedagogici
Orvosi Hetilap
Our Children
Our Schools Today

PCMR (President's Committee on Mental Retardation) Message
PTA Magazine
Pacesetters in Innovation
Pacific Medicine and Surgery
Pacific Sociological Review
Padiatrie und Padologie
Paedagogische Rundschau
Panminerva Medica
Papers in Psychology
Parents Magazine
Parents' Magazine and Better Family Living
Parents' Voice
Parks and Recreation
Past and Present
Pastoral Counselor
Pastoral Psychology
Pathologie et Biologie
Peabody Journal of Education
Pedagogia
Pedagogisk Forskning
Pedagogisk-Psychologisk Problem
Pediatric Clinics of North America
Pediatric Research
Pediatrics
Pediatriya
Pennsylvania Message
Pennsylvania Psychiatric Quarterly
Pennsylvania School Journal
Perception and Psychophysics
Perceptual and Motor Skills
Personnel
Personnel Administration
Personnel and Guidance Journal
Personnel Journal
Personnel Management
Personnel Management Abstracts
Personnel Practice Journal
Personnel Psychology
Perspectives in Biology and Medicine
Pflugers Archive-European Journal of Physiology
Pharmacological Reviews
Pharmakopsychiatrie Neuro-Psychopharmakologie
Phi Delta Kappan
Philosophical Review
Philosophy and Phenomenological Research
Philosophy of Science
Phylon
Physical Education
Physical Educator
Physical Therapy
Physiologia Bohemoslovenica
Physiology and Behavior
Pittsburgh Teachers Bulletin
Plastic and Reconstructive Surgery
Pointer
Polish Endocrinology
Polish Medical Journal
Population et Famille/Bevolking en Gezin
Postgraduate Medical Journal
Postgraduate Medicine
Practical Anthropology
Practica Oto-Rhino-Laryngologica
Praktische Psychologie
Praxis
Praxis der Kinderpsychologie und Kinderpsychiatrie
Praxis der Psychotherapie
Presse Medicale
Presspoints
Primates
Probleme und Ergebnisse der Psychologie
Proceedings of the Annual Convention of the American Psychological Association

Proceedings of the Annual Meeting of the Gerontological Society
Proceedings of the Indiana Academy of Science
Proceedings of the Invitational Conference on Testing Problems
Proceedings of the National Academy of Sciences, U. S.
Proceedings of the Royal Society of Medicine
Proceedings of the Society for Experimental Biology and Medicine
Proceedings of the Society for Psychical Research
Proceedings of the Southwestern Sociological Association
Programmed Instruction Bulletin
Programmed Learning
Programmed Learning and Educational Technology
Programs for the Handicapped
Progress in Cardiovascular Diseases
Progressive Architecture
Progressive Teacher
Project News of the Parsons State Hospital and Training School
Psicologia y Educacion
Psyche, Stuttgart
Psychedellic Review
Psychiatria Clinica
Psychiatria et Neurologia
Psychiatria et Neurologia Japonica
Psychiatria, Neurologia, and Neurochirurgia
Psychiatric Quarterly
Psychiatric Quarterly Supplement
Psychiatric Research Reports
Psychiatrie, Neurologie und Medizinische Psychologie
Psychiatry
Psychoanalytic Quarterly
Psychoanalytic Review
Psychologia Africana
Psychologia Africana Monograph Supplement
Psychologia: An International Journal of Psychology in the Orient
Psychologia a Patapsychologia Dietata
Psychologia Wychowawcza
Psychological Abstracts
Psychological Bulletin
Psychological Issues
Psychological Monographs
Psychological Record
Psychological Reports
Psychological Research Bulletin
Psychological Researches
Psychological Review
Psychologie Francaise
Psychologie und Praxis
Psychologie v Ekonomické Praxi
Psychologische Beiträge
Psychologische Forschung
Psychologische Praxis
Psychologische Rundschau
Psychology
Psychology in the Schools
Psychology Today
Psychometrika
Psychonomic Monograph Supplement
Psychonomic Science
Psychopharmacologia
Psychopharmacology Bulletin
Psychophysiology
Psychosomatic Medicine
Psychosomatics
Psychosynthesis Research Foundation
Psychotherapy and Psychosomatics
Psychotherapy: Theory, Research and Practice
Pszichologiai Tanulmányok
Public Administration
Public Administration Review
Public Health Reports
Public Health Service Publication
Public Interest
Public Opinion Quarterly
Public Personnel Review
Public Welfare
Purdue Opinion Panel Poll Report

Qawwi Qalbek

PUBLICATIONS SCANNED

Quarderni di Neuropsichiatria Infantile
Quarterly Journal of Experimental Psychology
Quarterly Journal of Medicine
Quarterly Journal of Speech
Quarterly Journal of Studies on Alcohol

Race
Radiation Research
Radiologia Clinica et Biologica
Radiology
Rajasthan University Studies
Rational Living
Reader's Guide
Reading Improvement
Reading Research Quarterly
Reading Teacher
Record
Recreation for the Handicapped
Recreation in Treatment Centers
Recreator
Reference Report (Washington State
Department of Institutions)
Reformed Review
Rehabilitation
Rehabilitation Counseling Bulletin
Rehabilitation in Australia
Rehabilitation Literature
Rehabilitation Record
Religion in Life
Religious Education
Remedial Education
Report from the Institute of Education,
U. Turku
Reports from the Psychological Institute,
U. Helsinki
Reports from the Psychological Laboratory,
University of Southern California
Reports of the Institute for Science of
Labour, Tokyo
Research Bulletin of the Department of
Psychology, Osmania U.
Research Bulletin of the National Institute
for Educational Research, Tokyo
Research in Education
Research Project, U. Canterbury
Research Quarterly
Research Quarterly of American Association
of Health, Physical Education, and Recreation
Research Report (Washington)
Research Reporter
Research Review (Washington State Department
of Institutions)
Restoration Quarterly
Review and Expositor
Review of Czechoslovak Medicine
Review of Educational Research
Review of Existential Psychology and
Psychiatry
Review of Religious Research
Revista Argentina de Psicologia
Revista Brasileira de Deficiencia Mental
Revista Brasileira de Estudos Pedagogicas
Revista de Etnografia si Folclor
Revista del Instituto de Ciencias Sociales
Revista de Neuro-Psiquiatria
Revista de Pedagogie, Rumania
Revista de Psicoanalisis
Revista de Psicologia General y Aplicada
Revista de Psicologia Normal e Patologica
Revista de Psicopatologia, Psicologia
Medica y Psicoterapia
Revista de Psihologie
Revista de Psiquiatria y Psicologia Medica
Revista de Statistics
Revista do Instituti Ciencias Sociaes da
Universidade do Brasil
Revista Interamericana de Psicologia
Revista Mexicana de Psicologia
Revista Mexicana de Sociologia
Revue de L'Universite d'Ottawa
Revue de Medecine Psychosomatique et de
Psychologie Medecale
Revue de Neuropsychiatrie Infantile et
d'Hygiene Mentale de l'Enfance
Revue de Psychologie Appliquee
Revue de Psychologie des Peuples

Revue des Ecoles
Revue D'Histoire Ecclesiastique
Revue d'Hygiene et de Medecine Sociale
Revue Francaise de Psychanalyse
Revue Francaise de Sociologie
Revue Internationale de Sociologie
Revue Neurologique
Revue Roumaine des Sciences Sociales%
Serie de Psychologie
Ricerca Scientifica
Ridge News, State Home and Training School,
Wheat Ridge, Colorado
Rivista Dell'Instuto Seroterapics
Italiana
Rivista di Psicologia della Scrittura
Rivista di Psicologia Sociale e Archivio
Italiano di Psicologia Generale e del
Lavoro
Rocky Mountain Social Science Journal
Romanian Medical Review
Royal Society of Medicine, Proceedings
Rural Sociology
Russkii Yazyk V Natsionalnoi (Shkole)

SK&F Psychiatric Reporter
Safety Education
Sak'art'velos SSR Mets'nierebat' Akademii
Moambe
Saturday Review
Sbornik Lekarsky
Sbornik Praci Filosoficke' Fakulty Brnenske'
University
Scandinavian Journal of Clinical and
Laboratory Investigation
Scandinavian Journal of Psychology
Scandinavian Studies
Scholastic Coach
Scholastic Teacher
School Activities
School Administration
School and Community
School and Society
School Arts
School Boards
School Counselor
School Guidance Worker
School Law Review
School Libraries
School Library Journal
School Lunch Journal
School Management
Schoolman's Week, University of Pennsylvania
School Musician Director and Teacher
School of Education Bulletin, Indiana
University
School Progress
School Review
School Safety
School Science and Mathematics
School Shop
Schule und Psychologie
Schweizer Erziehungs-Rundschau
Schweizerische Medizinische Wochenschrift
Science
Science and Society
Science Digest
Science Education
Science Journal
Sciences
Sciences Ecclesiastiques, Belgium
Science Teacher
Scientia Paedagogica Experimentalis
Scientific American
Scottish Educational Studies
Scottish Education Journal
Scottish Medical Journal
Securitas
Semaine des Hopitaux
Seminars in Psychiatry
Sight-Saving Review
Slow Learning Child
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Social and Clinical Psychology
Social Biology
Social Casework
Social Education

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- Social Forces
 Social Health News
 Social Problems
 Social Psychiatry
 Social Research
 Social Science
 Social Science and Medicine
 Social Science Information
 Social Science Quarterly
 Social Security Bulletin
 Social Service Review
 Social Studies
 Social Work
 Social Worker-Travailleur Social
 Sociologia, Brazil
 Sociological Abstracts
 Sociological Bulletin
 Sociological Inquiry
 Sociological Quarterly
 Sociological Review
 Sociologicky Casopis
 Sociology
 Sociology and Social Research
 Sociology of Education
 Sociometry
 Sotsiologiya VSSR, USSR
 South African Medical Journal
 Southern Education Report
 Southern Medical Bulletin
 Southern Medical Journal
 Southern Quarterly
 Southwestern Journal of Anthropology
 Southwestern Journal of Theology
 Sovetskaya Meditsina
 Sovetskaya Pedagogika
 Soviet Education
 Soviet Neurology and Psychiatry
 Soviet Psychology
 Soviet Review
 Soviet Sociology
 Sovremennye Problemy Deyatel'nosti i
 Stroeniya Tsentral'noi Nervnoi Sistemy
 Sozial Welt, Germany
 Special Education
 Special Education in Canada
 Special Education Review
 Spectrum
 Speech Monographs
 Speech Teacher
 Staff
 Steaua, Rumania
 Steroids
 Studia Psychologica
 Studi e Ricerche di Psicologia
 Studies and Research
 Studies in Art Education
 Studies in Higher Education
 Studies on the Left
 Surgery
 Surgery, Gynecology and Obstetrics
 Surgical Clinics of North America
- TR Times
 Tarbiz, Israel
 Teacher
 Teachers College Journal
 Teachers College Record
 Teaching Aids News
 Teaching and Training
 Teaching Exceptional Children
 Technical Bulletin of the Registry
 of Medical Technologists
 Techniques
 Tennessee Teacher
 Teoriija i Praktika Fizicheskoi Kulitury
 Teratology
 Texas Medicine
 Texas Outlook
 Texas Reports on Biology and Medicine
 Theology and Life
 Theology Today
 Theoria: A Swedish Journal of Philosophy
 Theory into Practice
 Thorax
 Tidsskrift for den Norske Laegeforening, Oslo
 Tijdschrift voor Geneeskunde, Louvain
- Tijdschrift voor Zwakzinnigheid en
 Zwakinnigenzorg
 Times Educational Supplement, London
 Today's Child
 Today's Education
 Tohoku Journal of Experimental Medicine
 Tohoku Psychologica Folia
 Toronto Education Quarterly
 Traffic Safety Research Review
 Training School Bulletin
 Traite' de Psychologie Experimentale
 Trans-Action
 Transactions of the New York Academy of
 Sciences
 Transactions of the Westmarck Society
 Transfusion
 Transplantation
 Travail Humain
 Trends in Education
 Trudy Leningradskogo Nauchno-Issledovatel'
 skogo Instituta Ekspertizy Trudospobob-
 nosti i Organizatsii Truda Invalidov
 Trustee
- UNESCO Courier
 Ugeskrift for Laeger, Copenhagen
 Union Medicale du Canada
 United States Office of Education
 Publications
 Universities Quarterly
 Urban Education
 Urban Reporter
 Urban Review
 Urban Studies
 Urim
 Utah Educational Review
- V.O.C. Journal of Education
 Vascular Diseases
 Vestnik Akademii Meditsinskikh Nauk SSSR
 Vestnik Akademii Nauk SSSR
 Vestnik Oftalmologii, Moscow
 Vestnik Otorinolaringologii, Moscow
 Vestnik Vysshei Shkoly
 Viata Romineasca
 Victoria University of Wellington
 Publications in Psychology
 Virginia Journal of Education
 Virology
 Visual Communications Instructor
 Visual Education
 Vocational Guidance Quarterly
 Voix Silence
 Volta Review
 Volunteer Council News (Lynchburg Training
 School and Hospital)
 Voprosy Filosofii
 Voprosy Psikhologii
 Voprosy Sravnitel'noi Fiziologii Analizatorov
 Vox Sanguinis
 Vrachebnoe Delo, Kiev
- WHO (World Health Organization) Bulletin
 WHO (World Health Organization) Chronicle
 Warren G. Murray Children's Center (State
 of Illinois Department of Mental Health)
 Washington Education
 Washington University Department of
 Psychology Technical Report
 Welfare in Review
 Welfare Reporter
 Welt der Schule
 Wesleyan Studies in Religion
 West Virginia School Journal
 Wiener Klinische Wochenschrift
 Wiener Medizinische Wochenschrift
 Wilson Library Bulletin
 Winnower
 Wisconsin Journal of Education
 Works of the Institute of Higher Nervous
 Activity: Pathophysiological Series
 World Year Book of Education
- Yale Journal of Biology and Medicine

PUBLICATIONS SCANNED

Young Children

Zeitschrift Evangelische Ethik
 Zeitschrift fuer Kirchengeschichte
 Zeitschrift fur Altersforschung
 Zeitschrift fur die Gesamte Innere Medizin
 und ihre Grenzgebiete
 Zeitschrift fur Experimentelle und
 Angewandte Psychologie
 Zeitschrift fur Heilpädagogik
 Zeitschrift fur Kinderheilkunde, Berlin
 Zeitschrift fuer Paedagogik

Zeitschrift fur Psychologie
 Zeitschrift fur Psychosomatische Medizin
 und Psychoanalyse
 Zeitschrift fur Psychotherapie und
 Medizinische Psychologie
 Zeitschrift fur Theologie und Kirche
 Zeitschrift fur Tierpsychologie
 Zentralblatt fur Chirurgie
 Zentralblatt fur Gynakologie
 Zeszyty Naukow
 Zhurnal Nevropatologii i Psikhatrii
 Zhurnal Obshchei Biologii
 Zhurnal Vyshei Nervnoi Deyatel'nost:
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Abbreviations:

b.i. brain injury, brain injured
CNS central nervous system
CP cerebral palsy
c/w compared with
EMR educable mentally retarded
ep. epilepsy
fr from

inst. institution, institutionalized
MR mentally retarded
PMR profoundly mentally retarded
SMR severely mentally retarded
spec. ed. special education
TMR trainable mentally retarded
w/ with

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